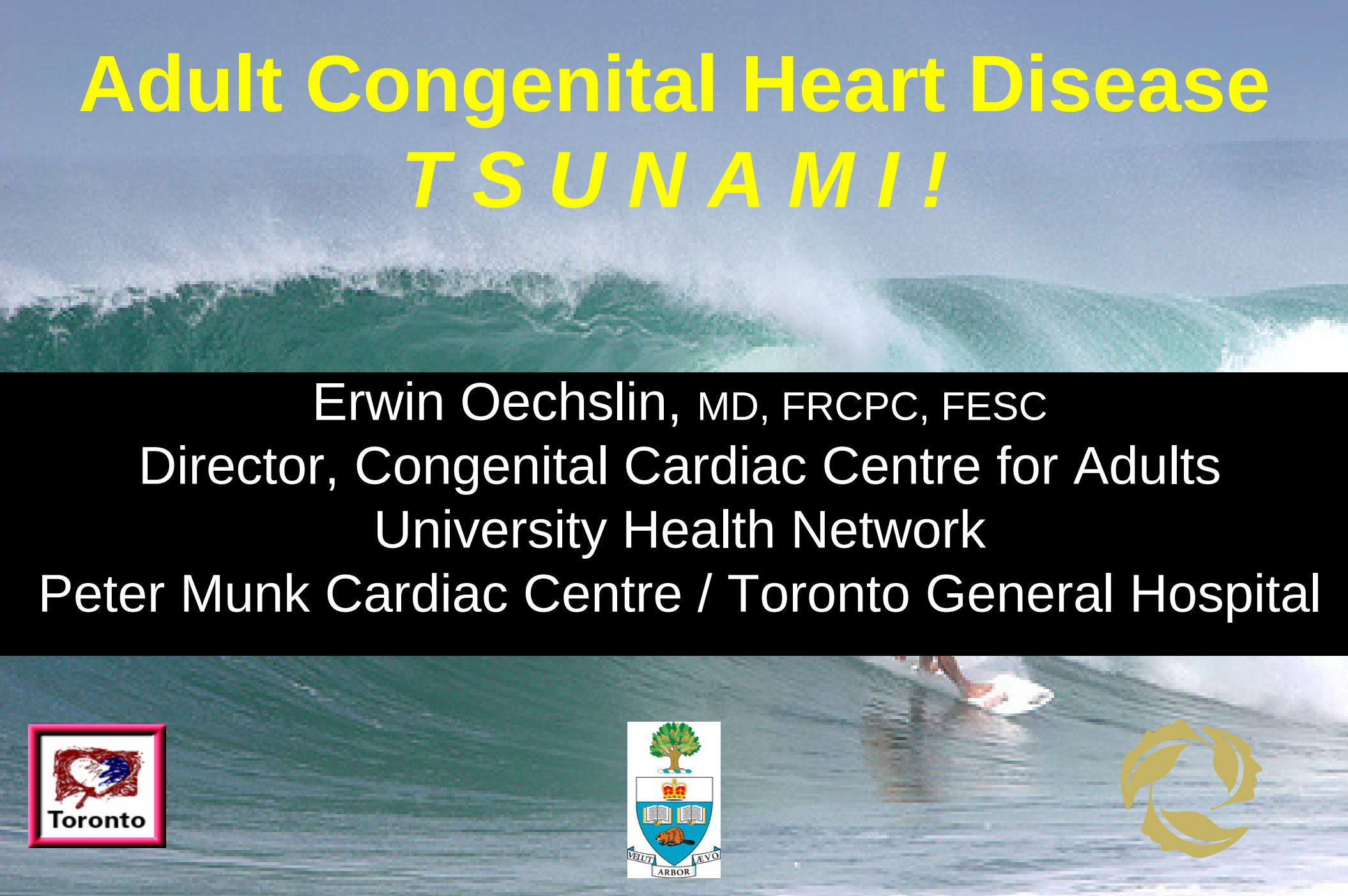
Adult Congenital Heart Disease

T S U N A M I !

Erwin Oechslin, MD, FRCPC, FESC
Director, Congenital Cardiac Centre for Adults
University Health Network
Peter Munk Cardiac Centre / Toronto General Hospital



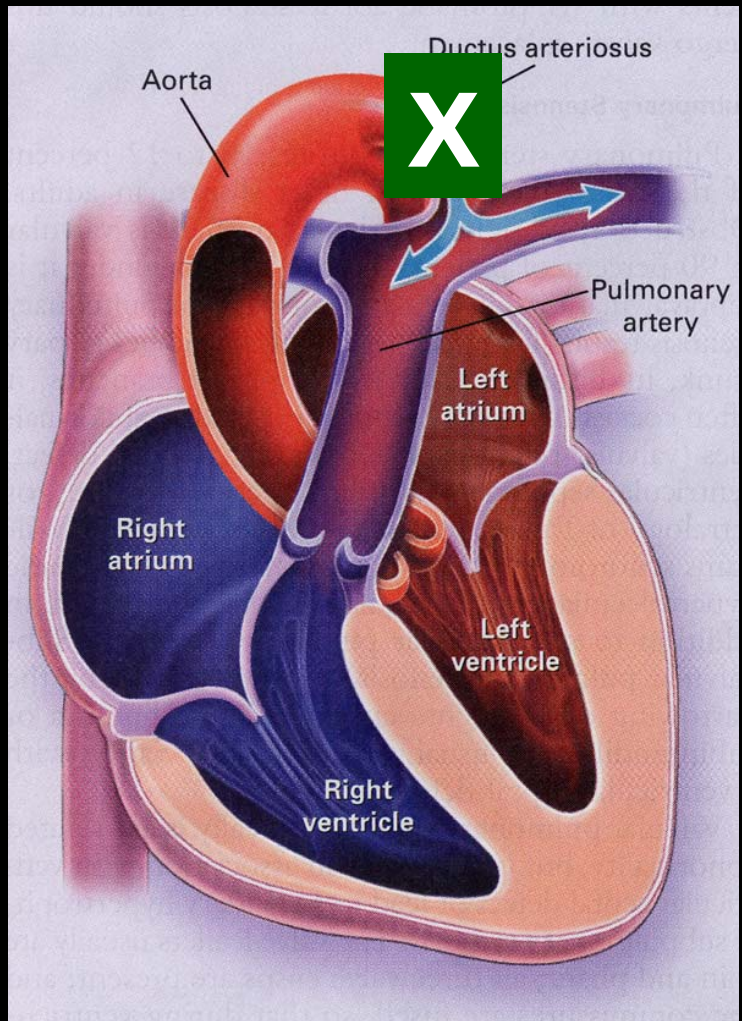
Outline

- History
- The adult population
- Do we cure CHD patients?
- Summary

1938

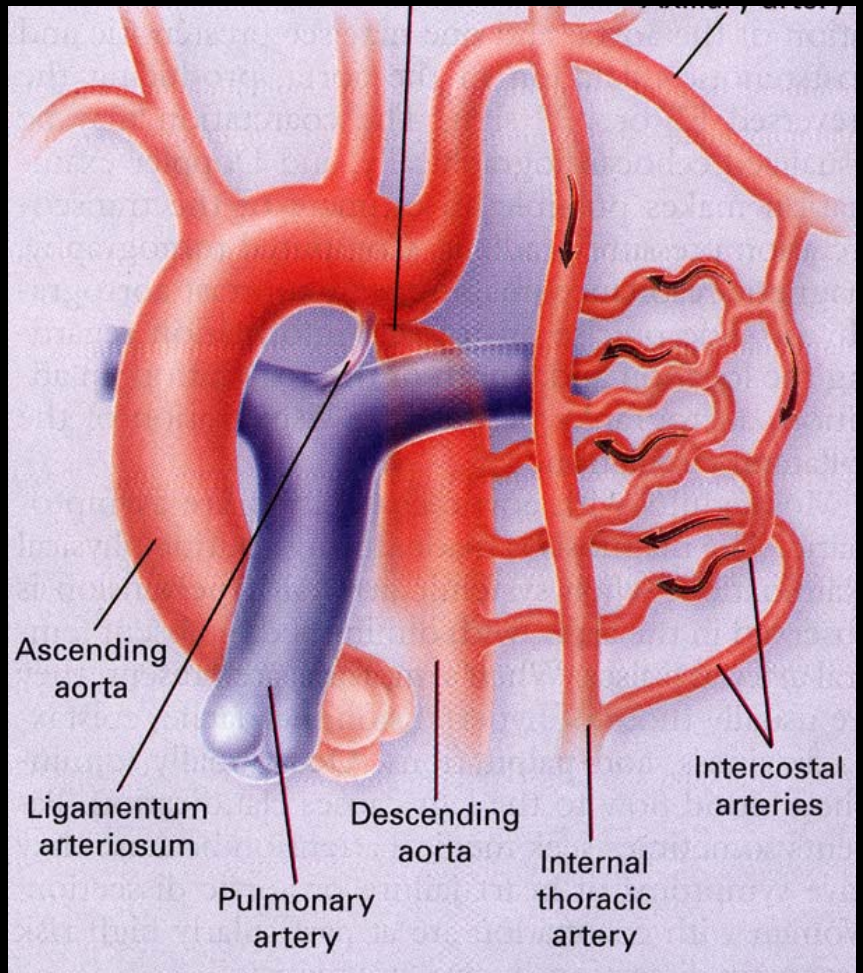
1st Operation in CHD

R.E. Gross
Boston, USA

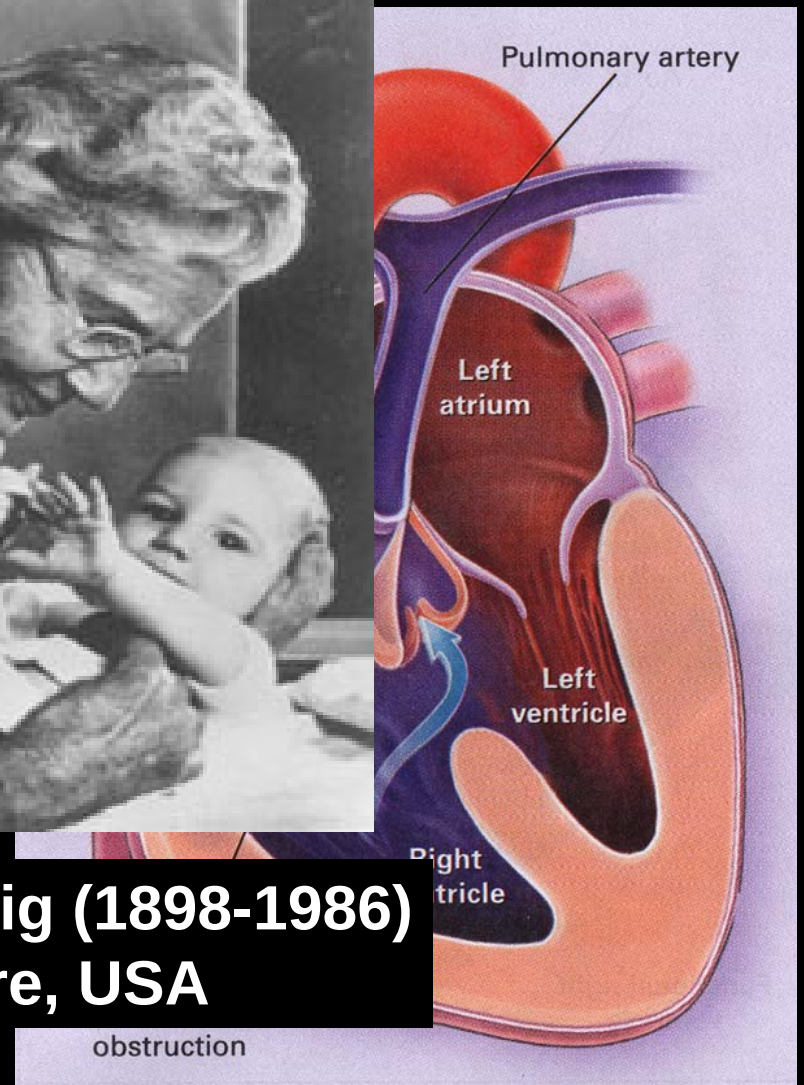


1944 - Operation Aortic Coarctation

C. Crafoord
Stockholm, SE

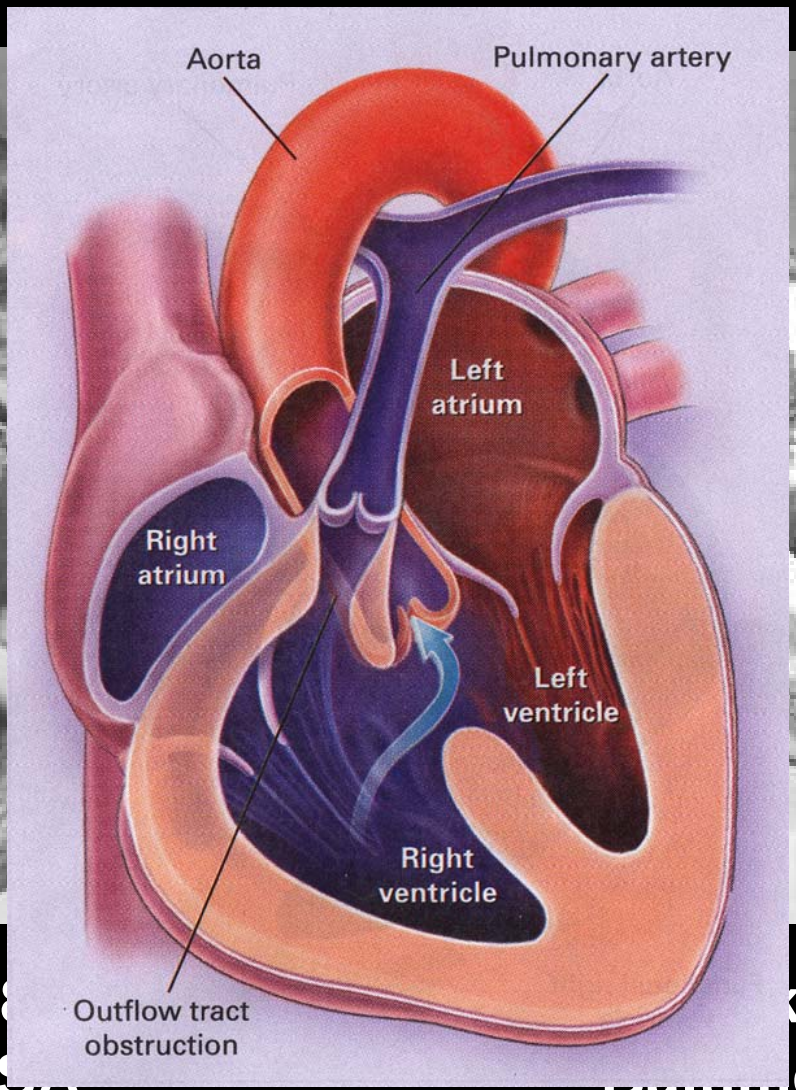


Tetralogy of Fallot



Helen B. Taussig (1898-1986)
Baltimore, USA

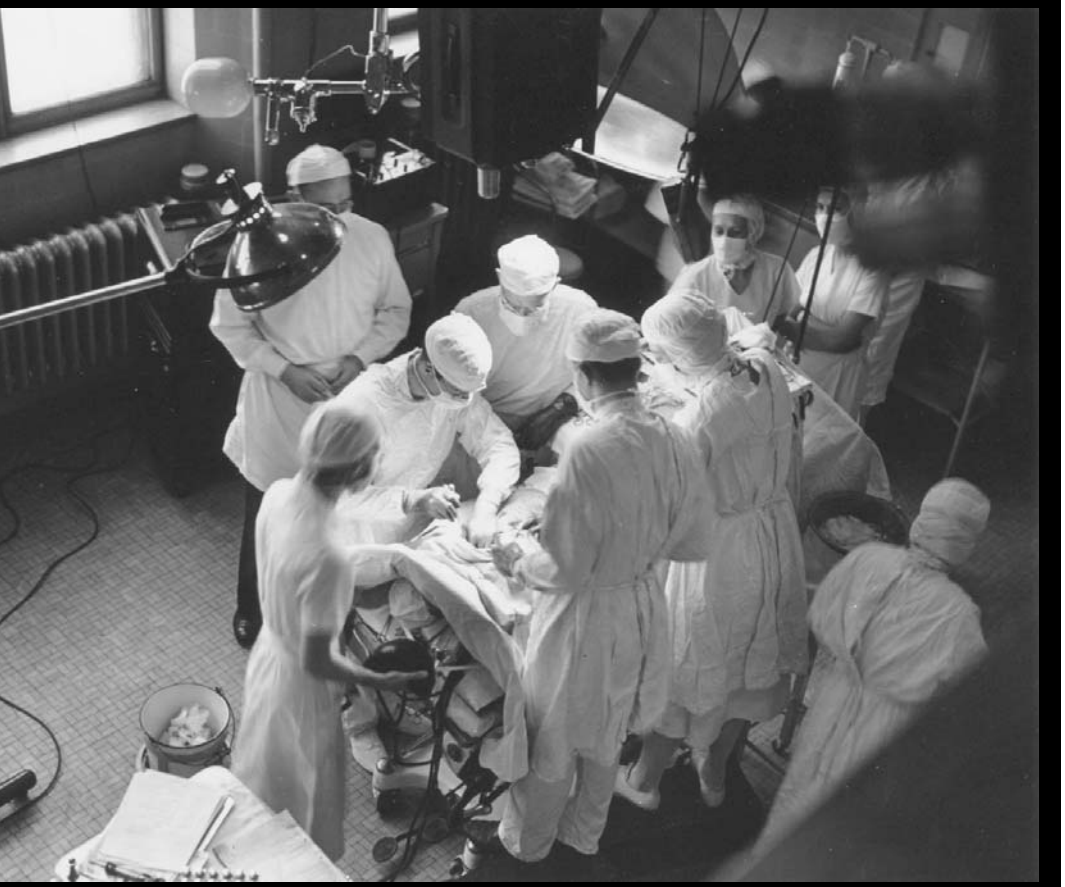
1944



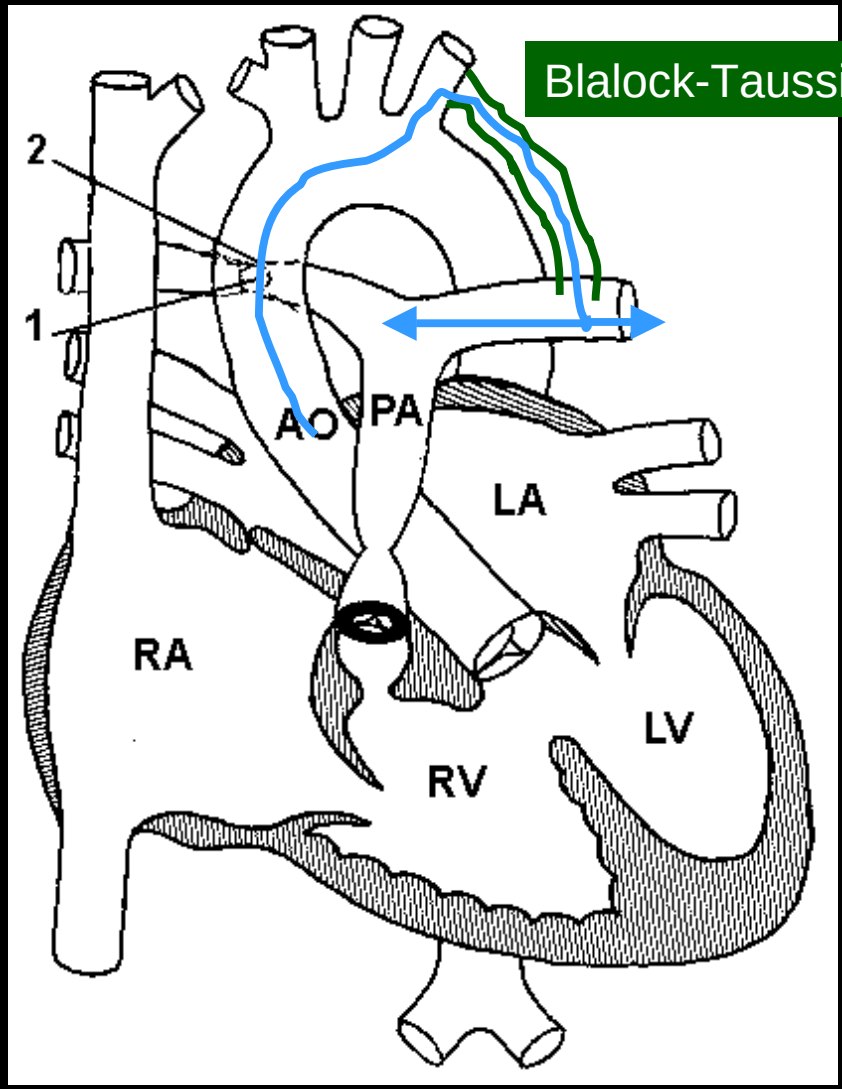
Helen B. Taussig (18
Baltimore, USA

(1899-1964)
Baltimore, USA

Tetralogy of Fallot



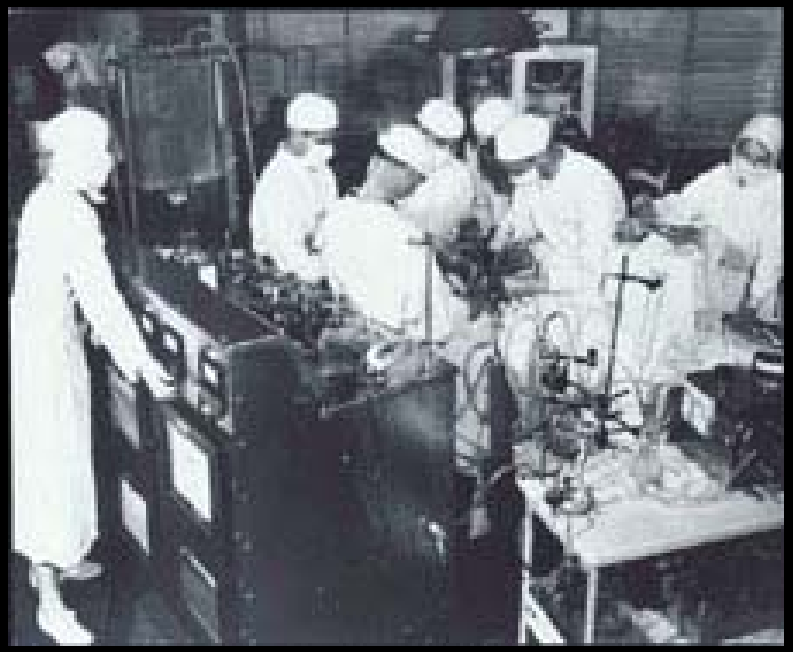
Dr. A. Blalock and
his team



Alfred Blalock
1899-1964

Vivien Thomas
1910-1985

1st Heart Lung Machine in 1953

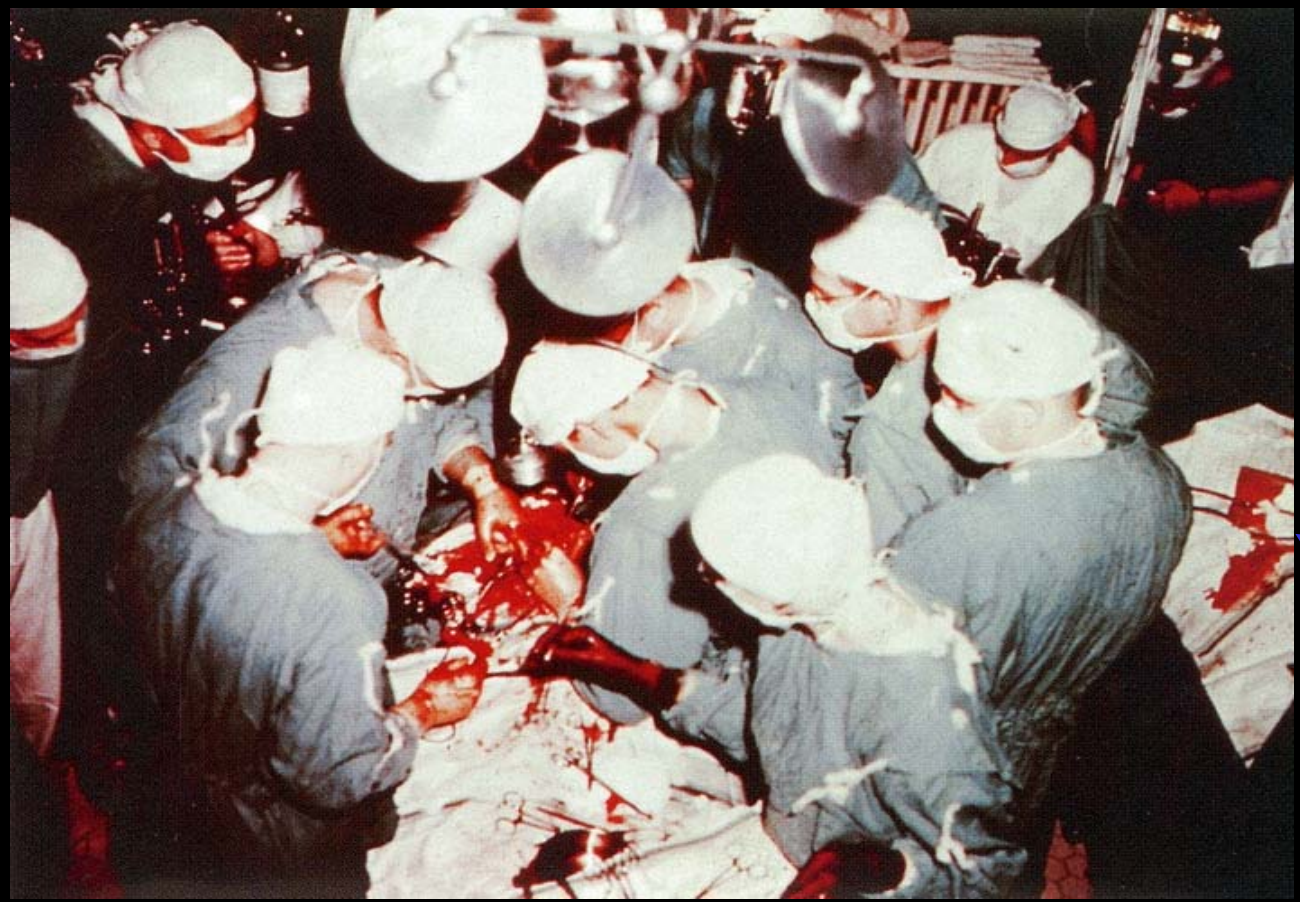


John H. Gibbon Jr. 1903 - 1973

- **Open Heart Surgery**
- **Intracardiac Repair**

C.W. Lillehei, Minneapolis, USA: First Tetralogy of Fallot Repair

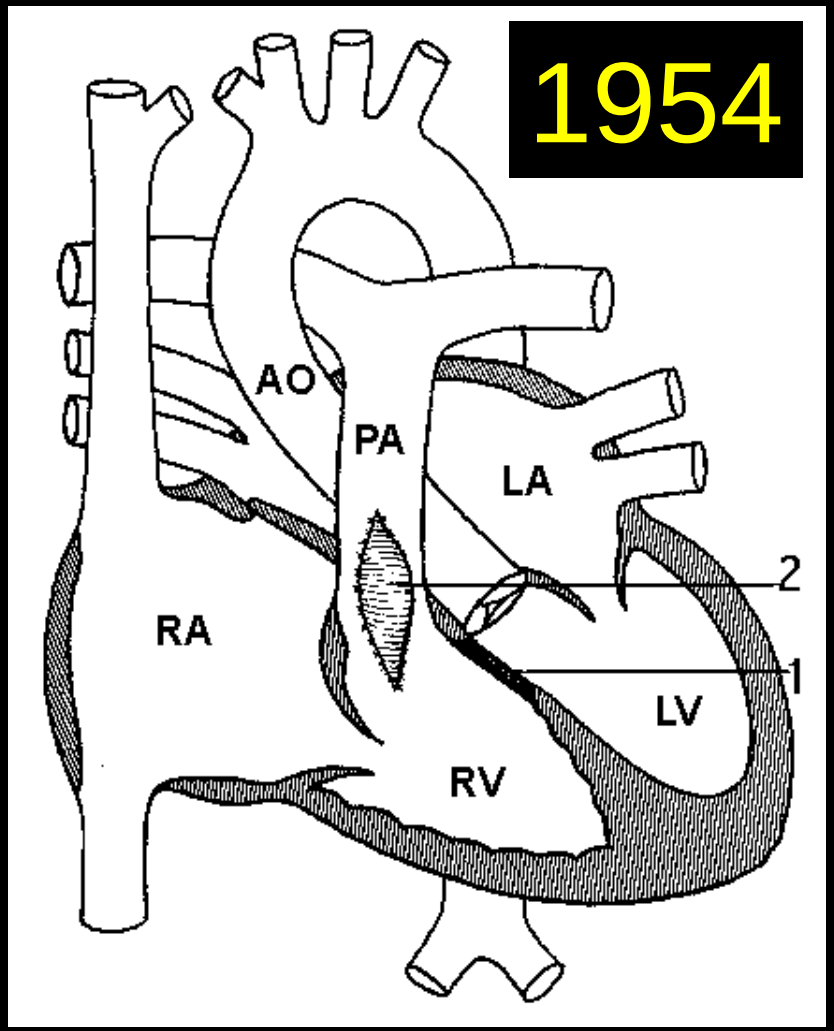
August 31, 1954



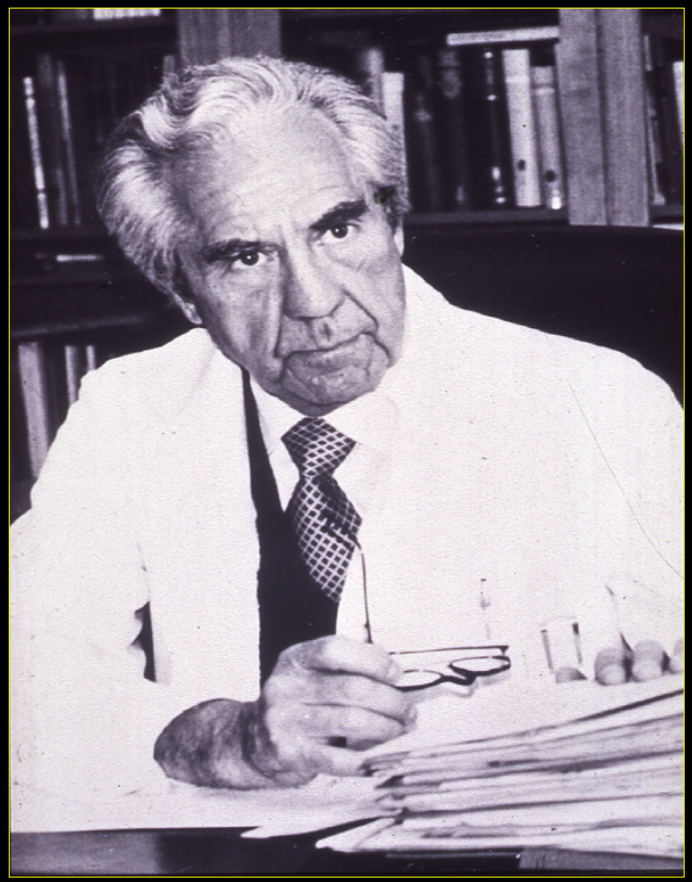
Cross-Circulation

Courtesy of Dr. W. Williams, Toronto

C.W. Lillehei, Minneapolis, USA: First Tetralogy of Fallot Repair

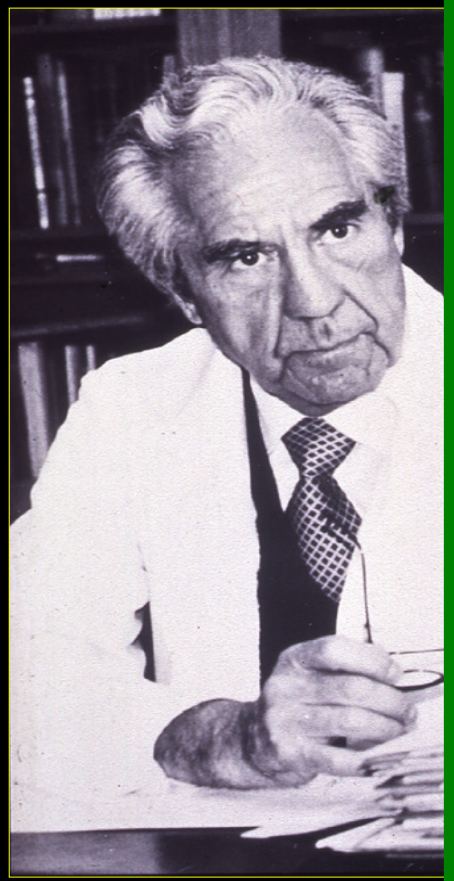


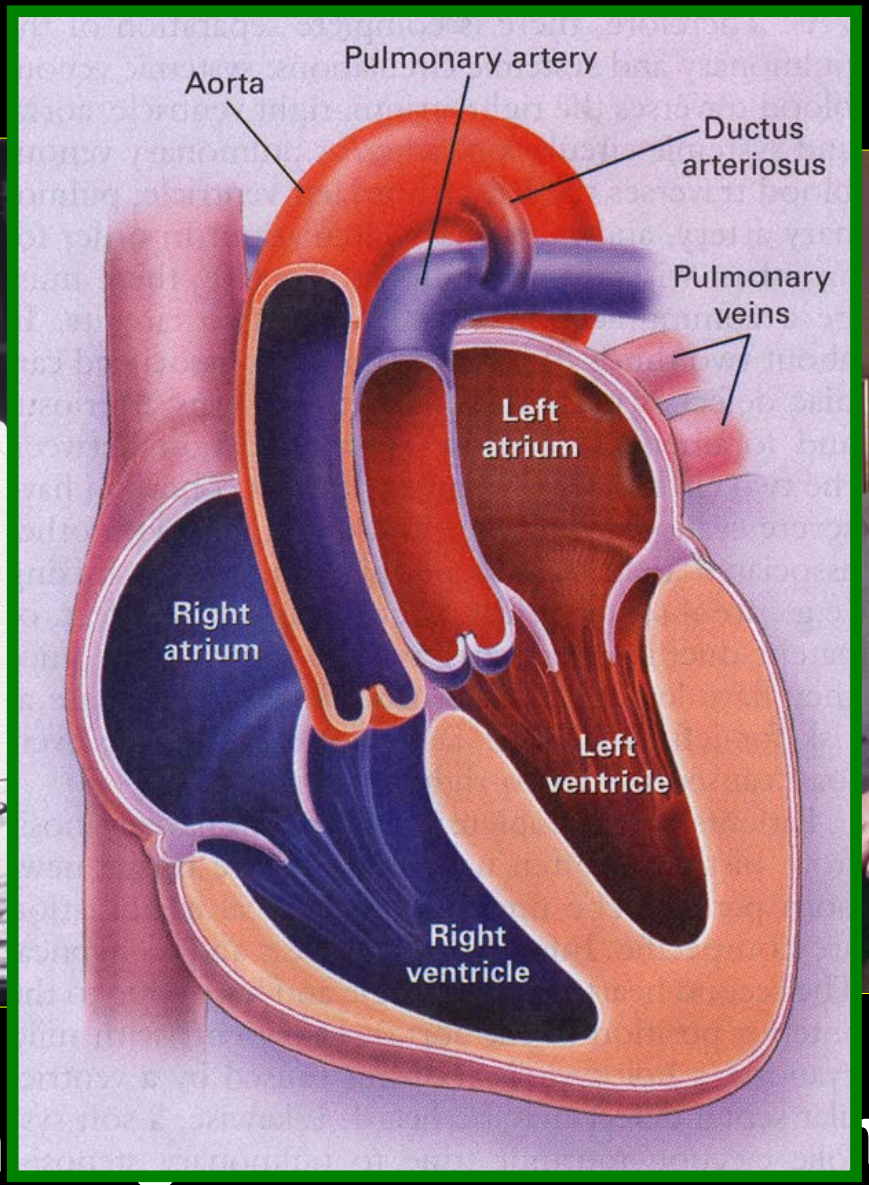
- VSD patch closure
- RVOT reconstruction
  - Transannular patch (fig.)
  - Valvotomy, infundibulectomy, RV patch enlargement
  - RV-PA conduit

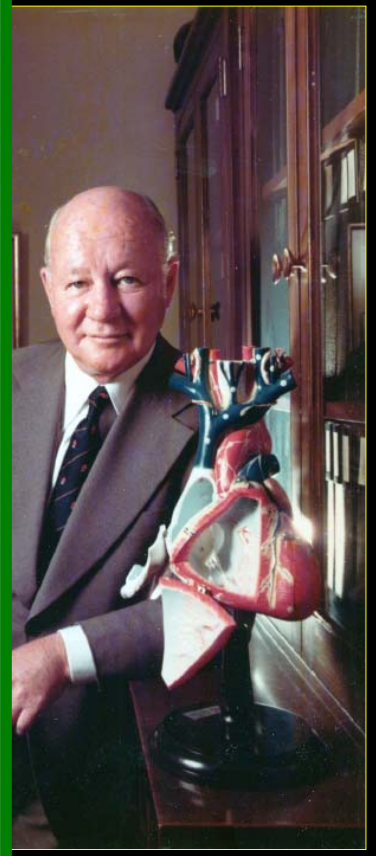


Åke Senning
Zürich, CH

William T. Mustard
Toronto, Ca

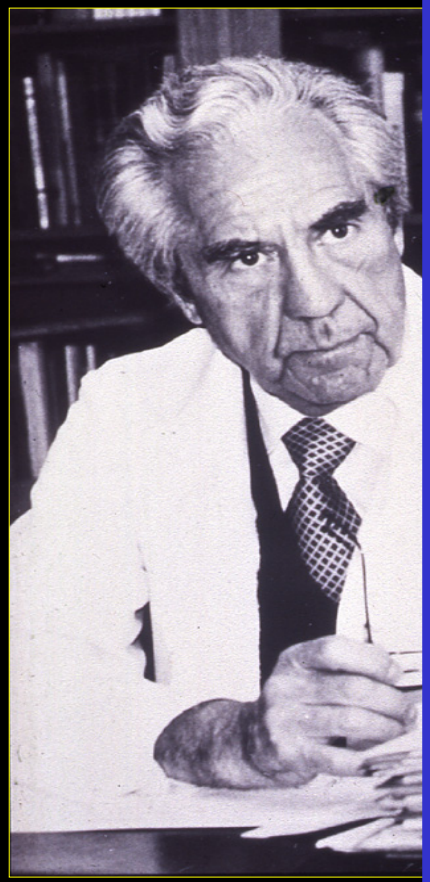


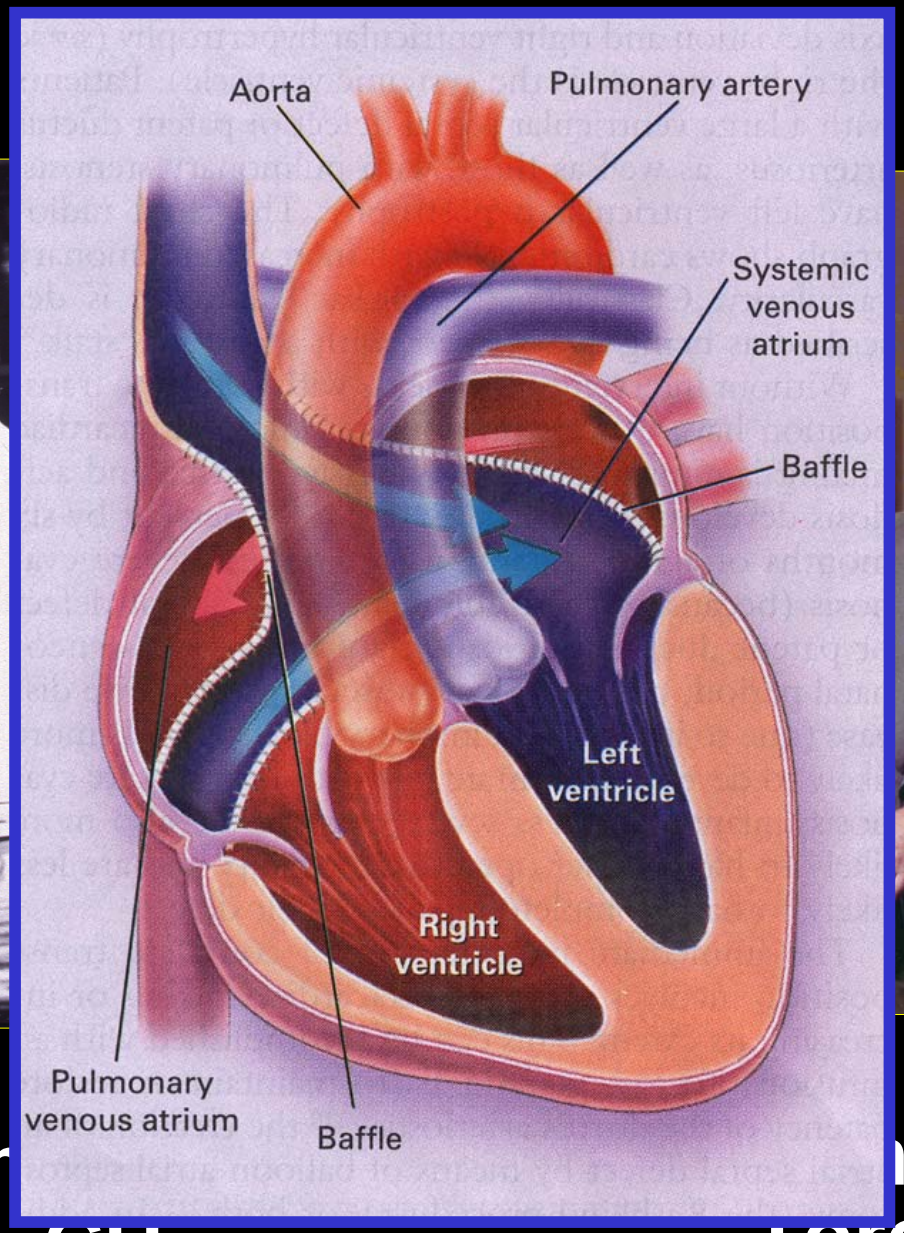


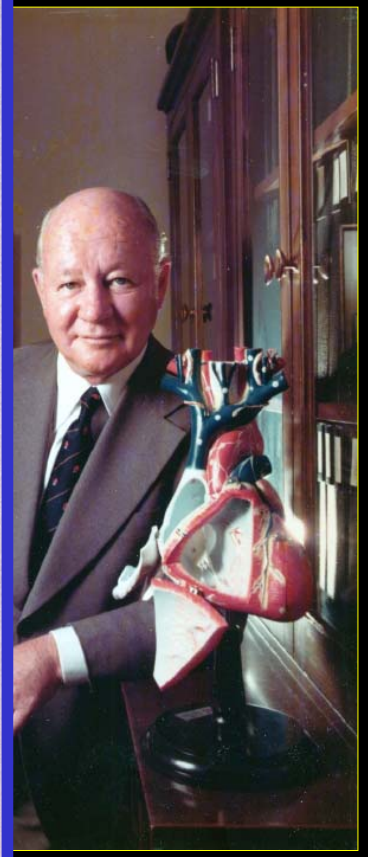


Åke Sen
Zürich, CH

n T. Mustard
Toronto, Ca

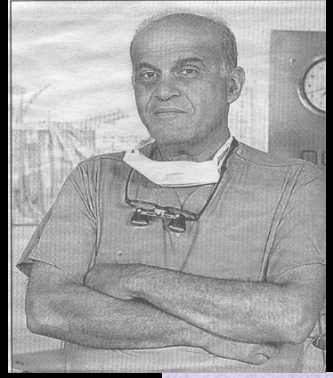




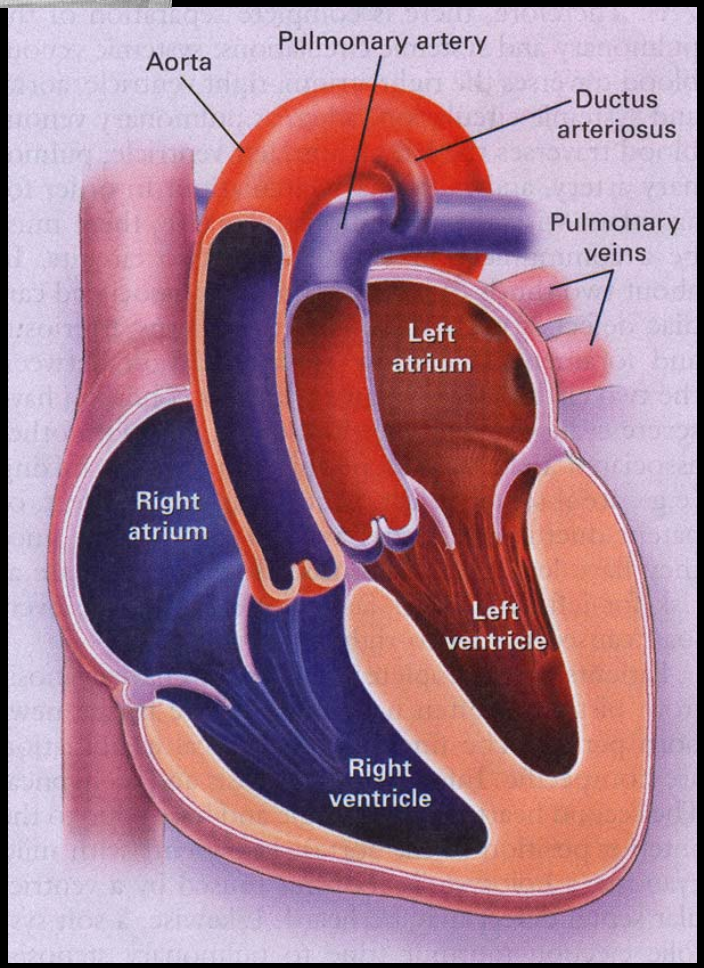


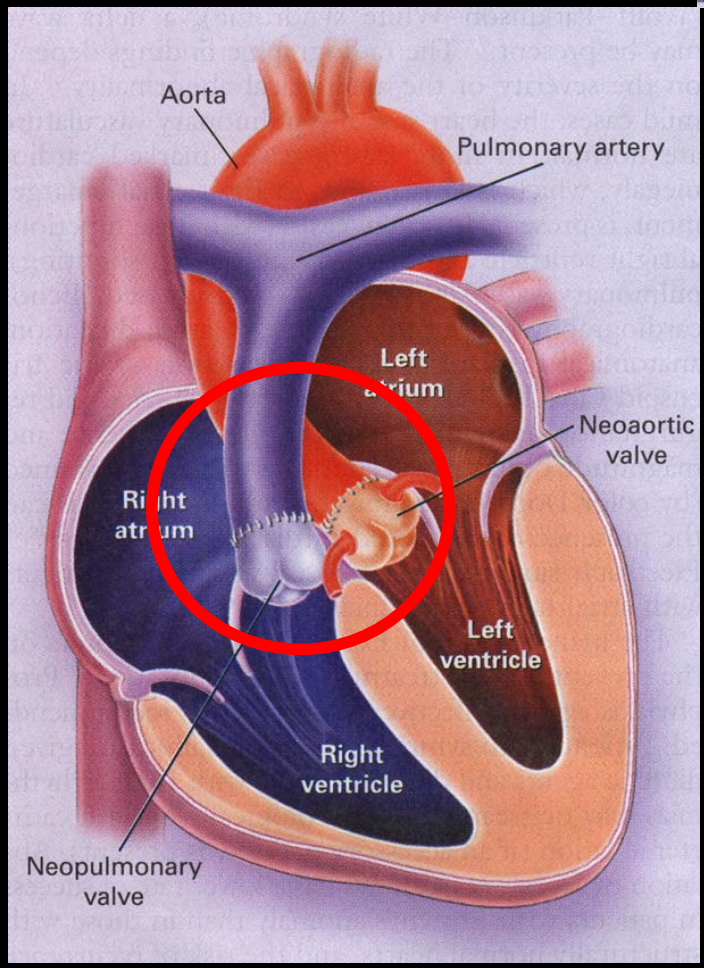
Åke Ser
Zürich, CH

T. Mustard
Toronto, Ca

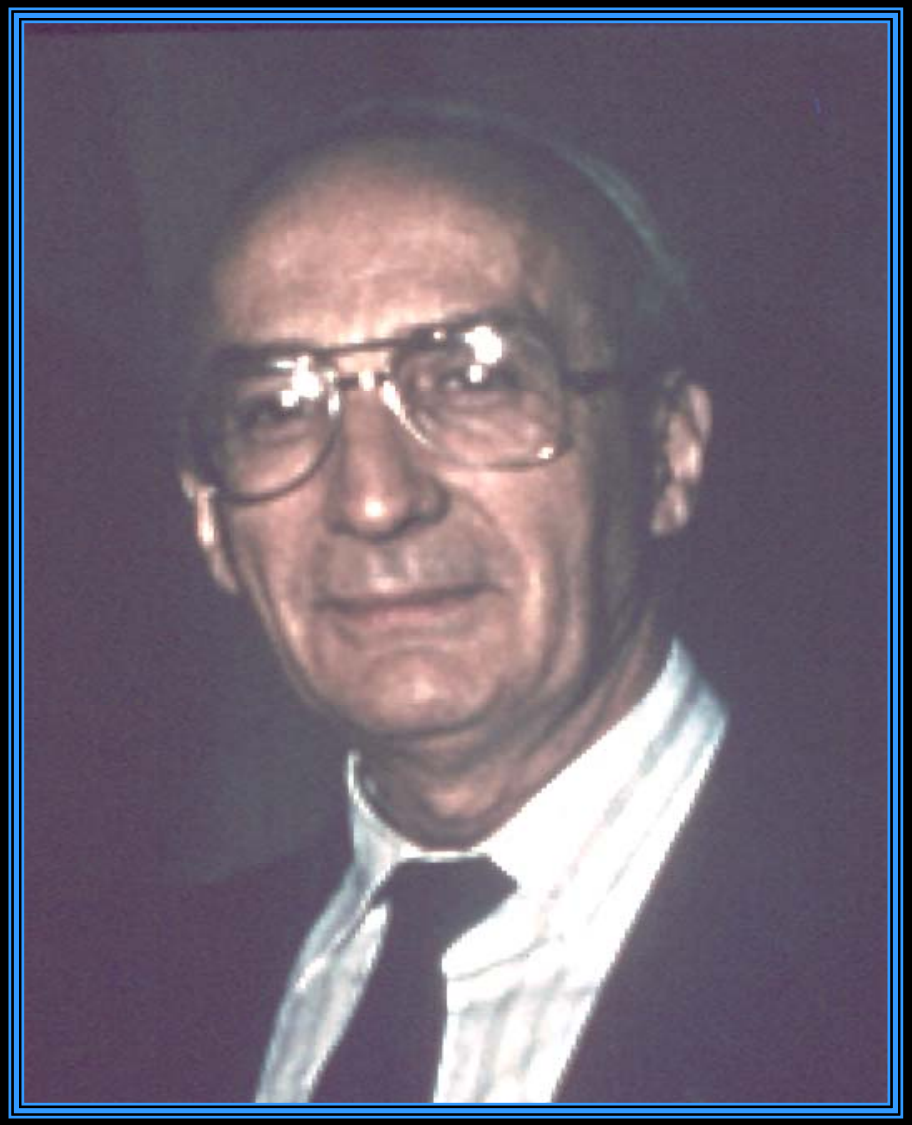


1975 - *ARTERIAL* Switch Procedure



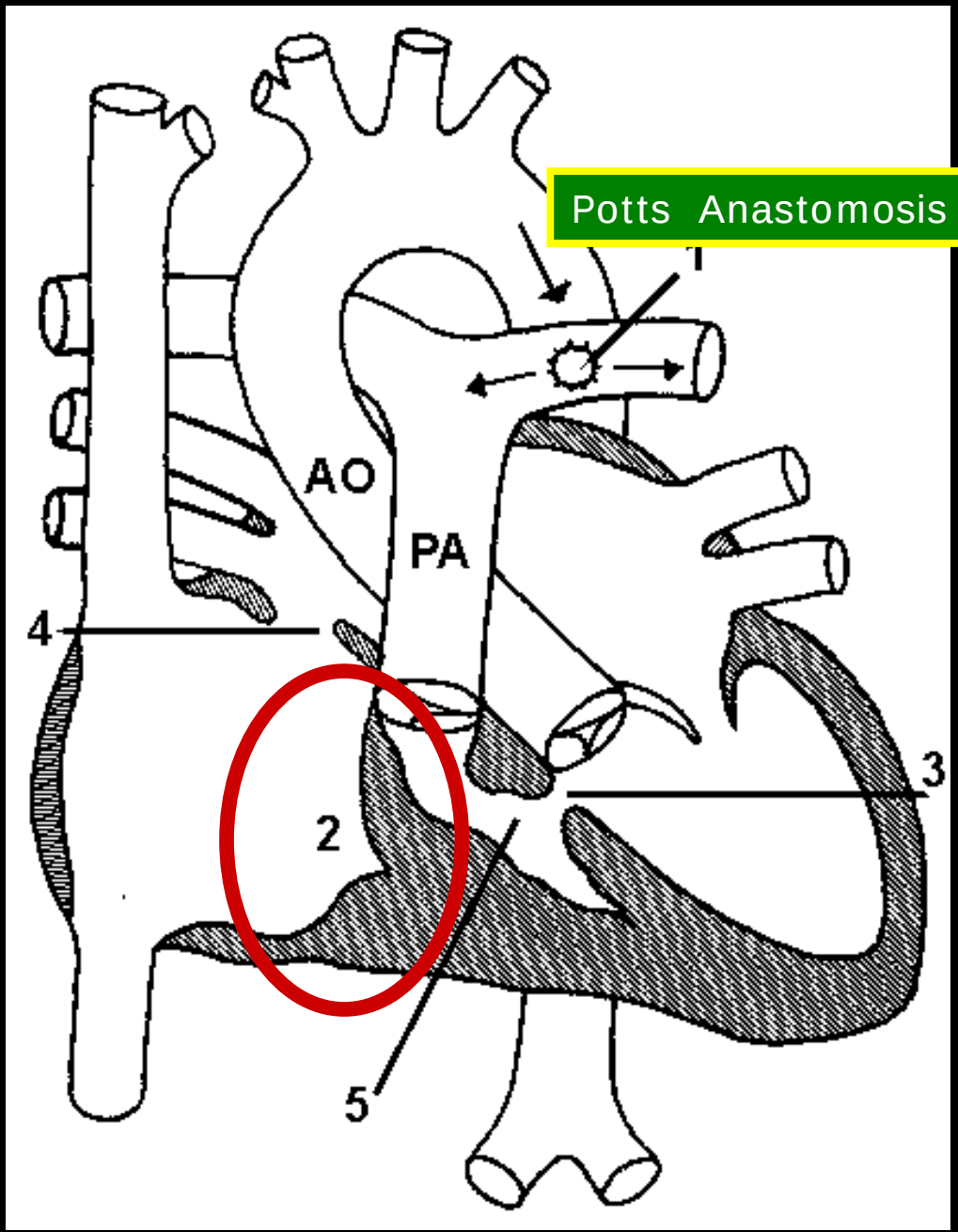


Brickner ME, et al. N Engl J Med 2000;342:334-342



F. Fontan

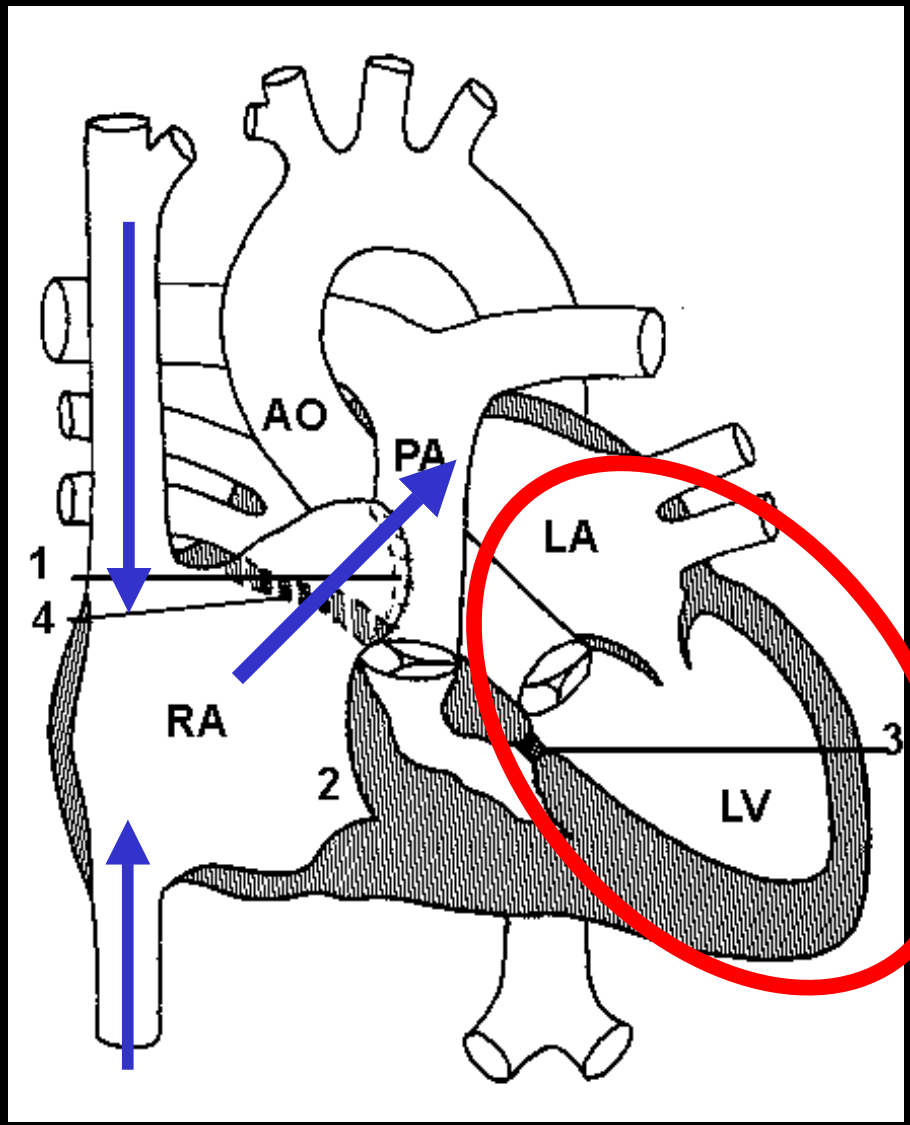
Tricuspid Atresia with ASD and VSD



Fontan Procedure

- Not suitable for biventricular repair
- Separation of the systemic and pulmonary circulation

1971: Fontan - Circulation



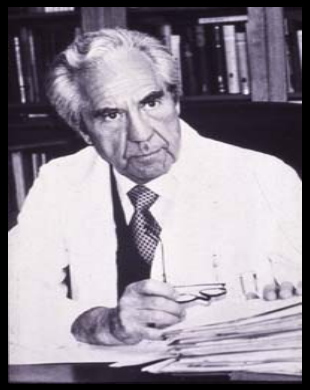
Gross

Crafoord

Taussig

Blalock

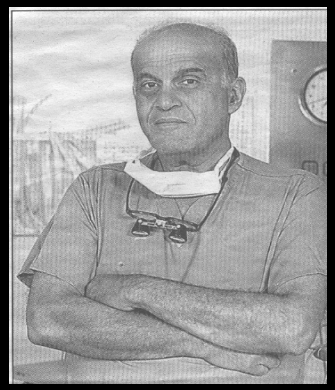
Lillehei



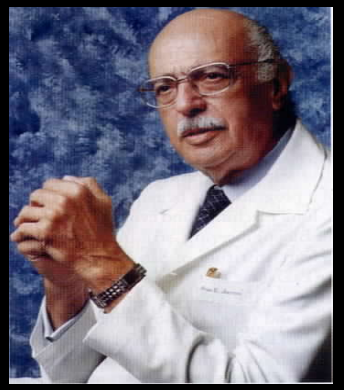
Senning

Mustard

Fontan

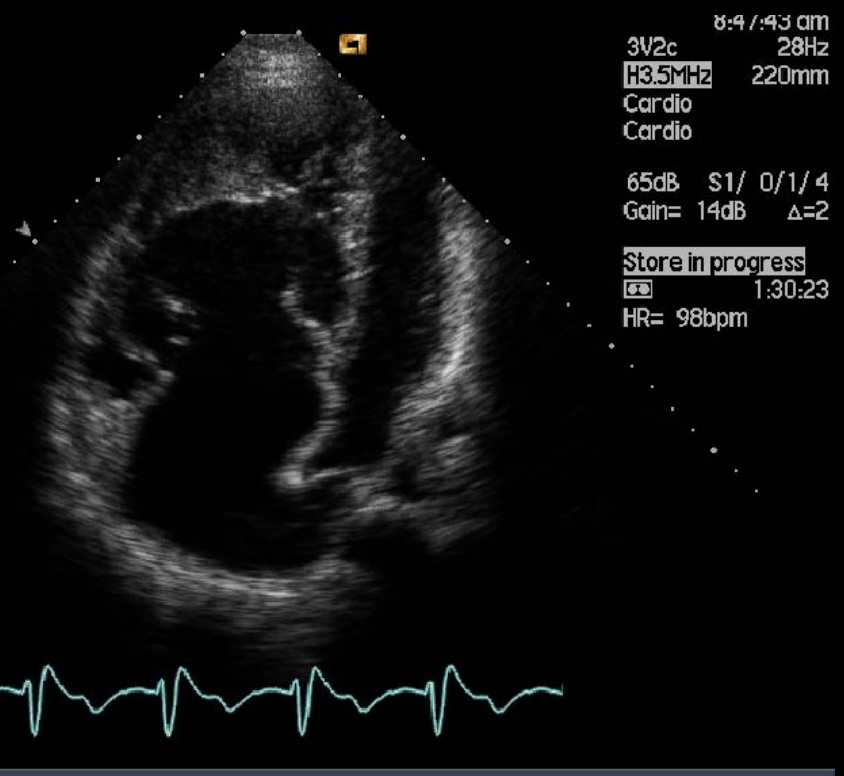


Yacoub

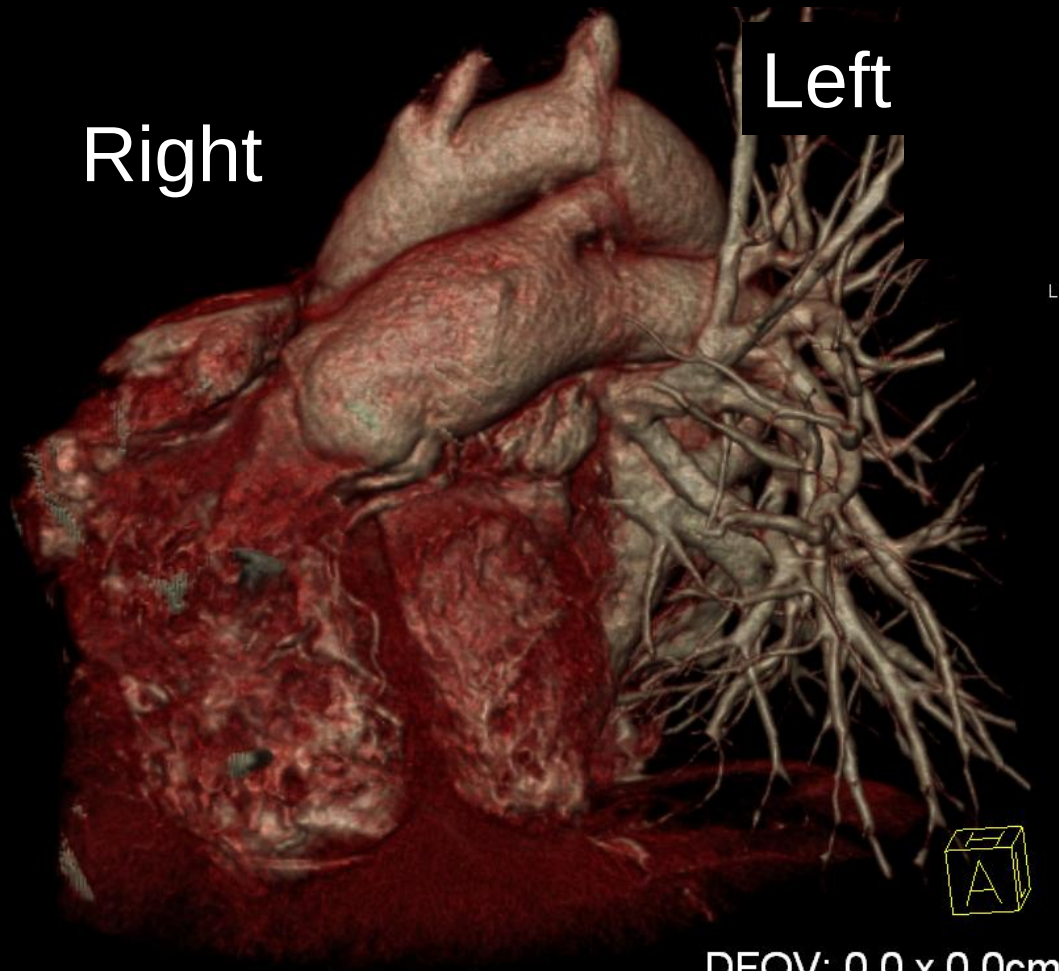


Jatene

Diagnostic Imaging



0.0 kV
0.0 mA
Tilt: 0.0
0.0 s
W:255 L:127

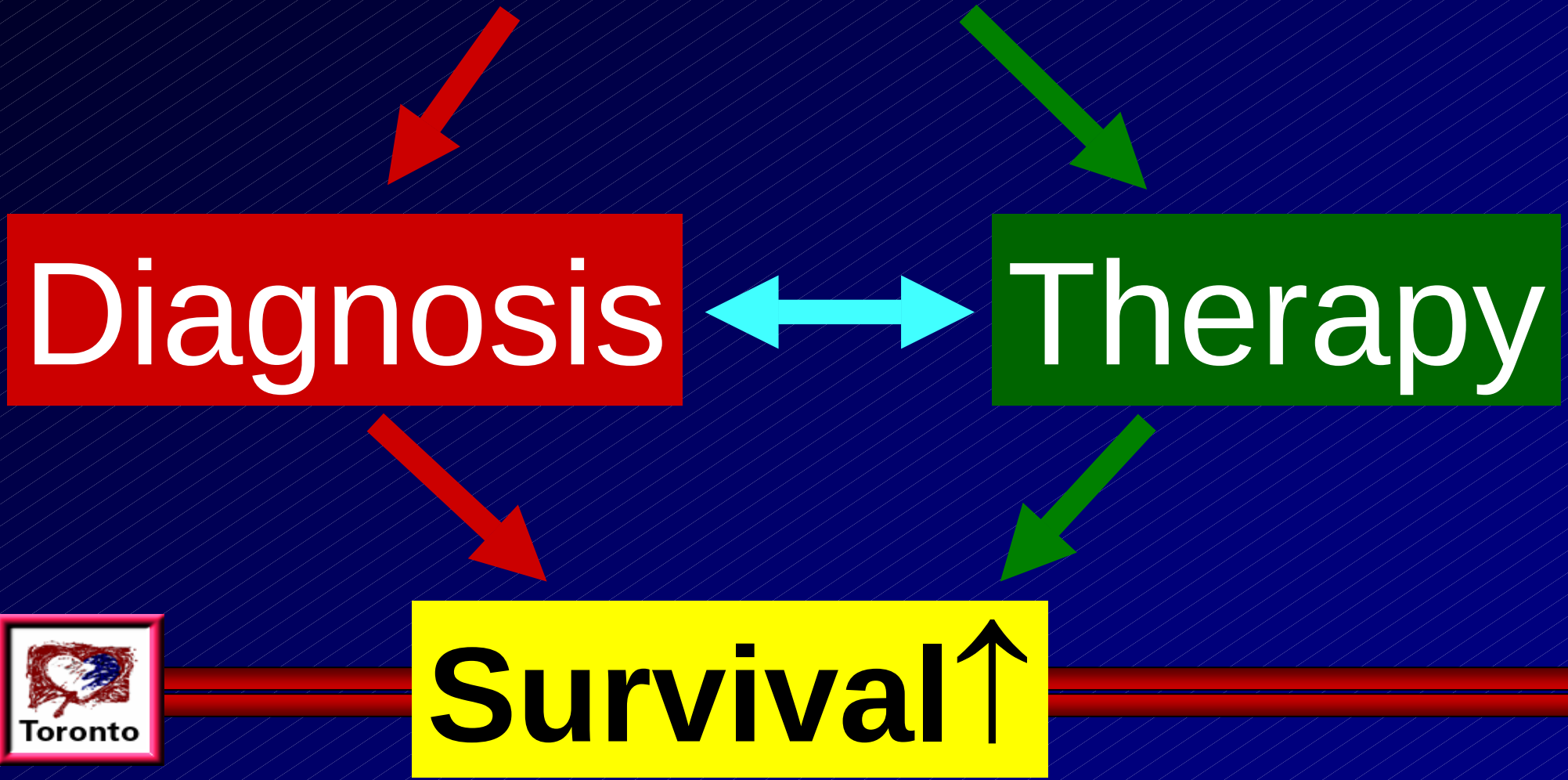


DFOV: 0.0 x 0.0cm

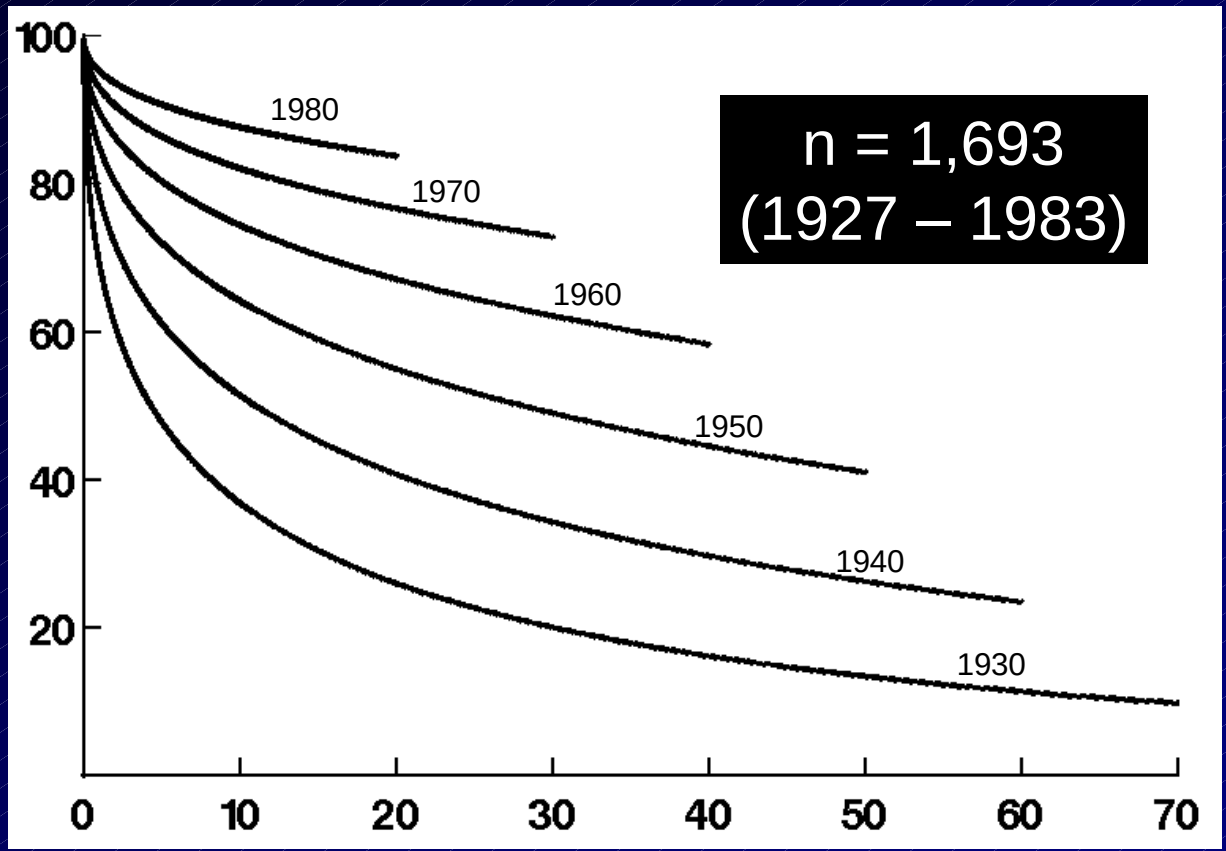
Toronto CHD History

- Hospital for Sick Children
  - Keith, Rowe, Freedom, Redington
  - Mustard, Trusler, Williams, Van Arsdell
- ACHD clinic opened 1959 at Toronto General Hospital

Advances



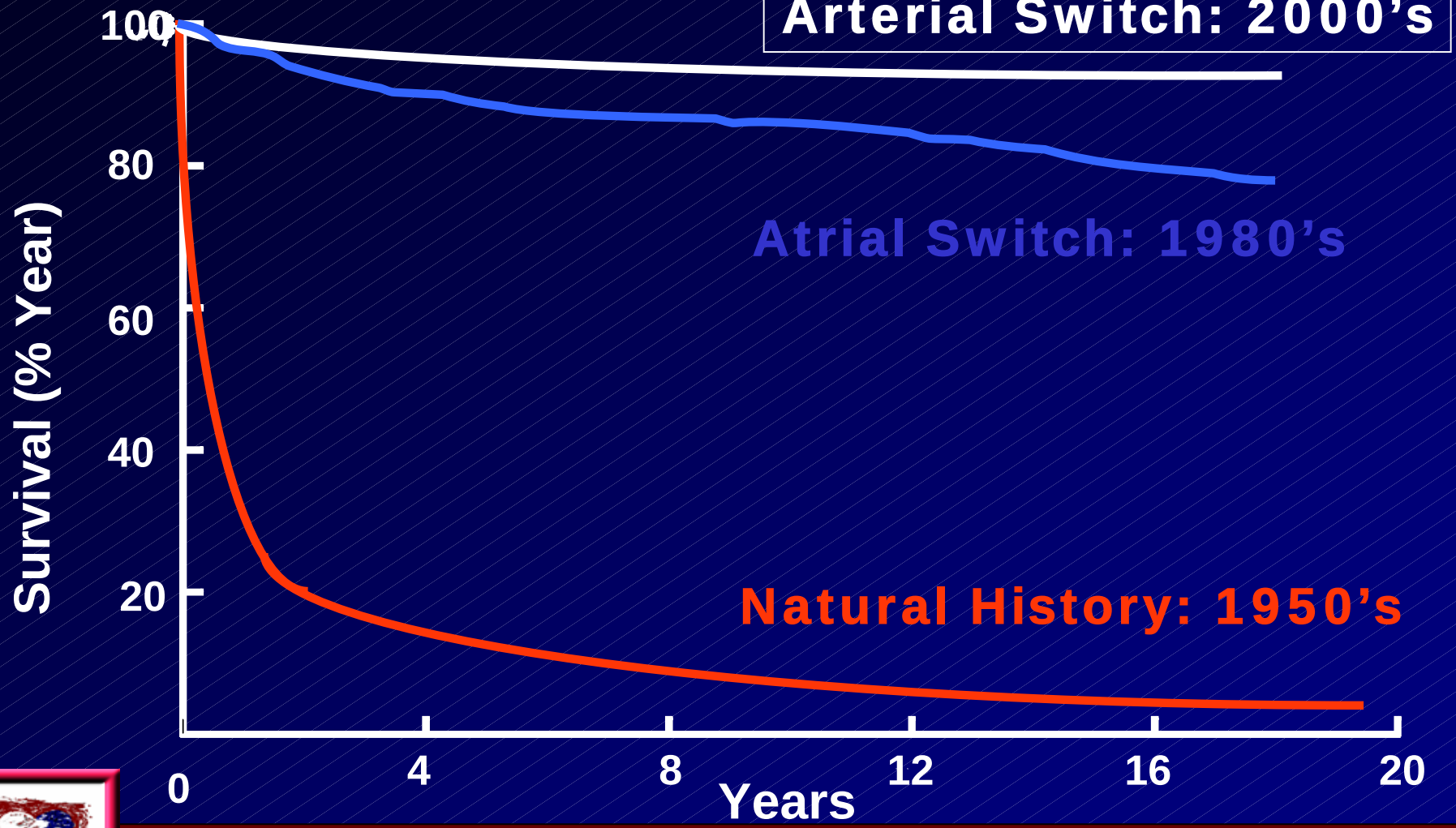
Survival in Tetralogy of Fallot by decade of birth at HSC, Toronto



Toronto

Hickey EJ, Manlhiot C, McCrindle BW 2007
Heart and Stroke Foundation of Ontario “Functional outcomes in Tetralogy of Fallot

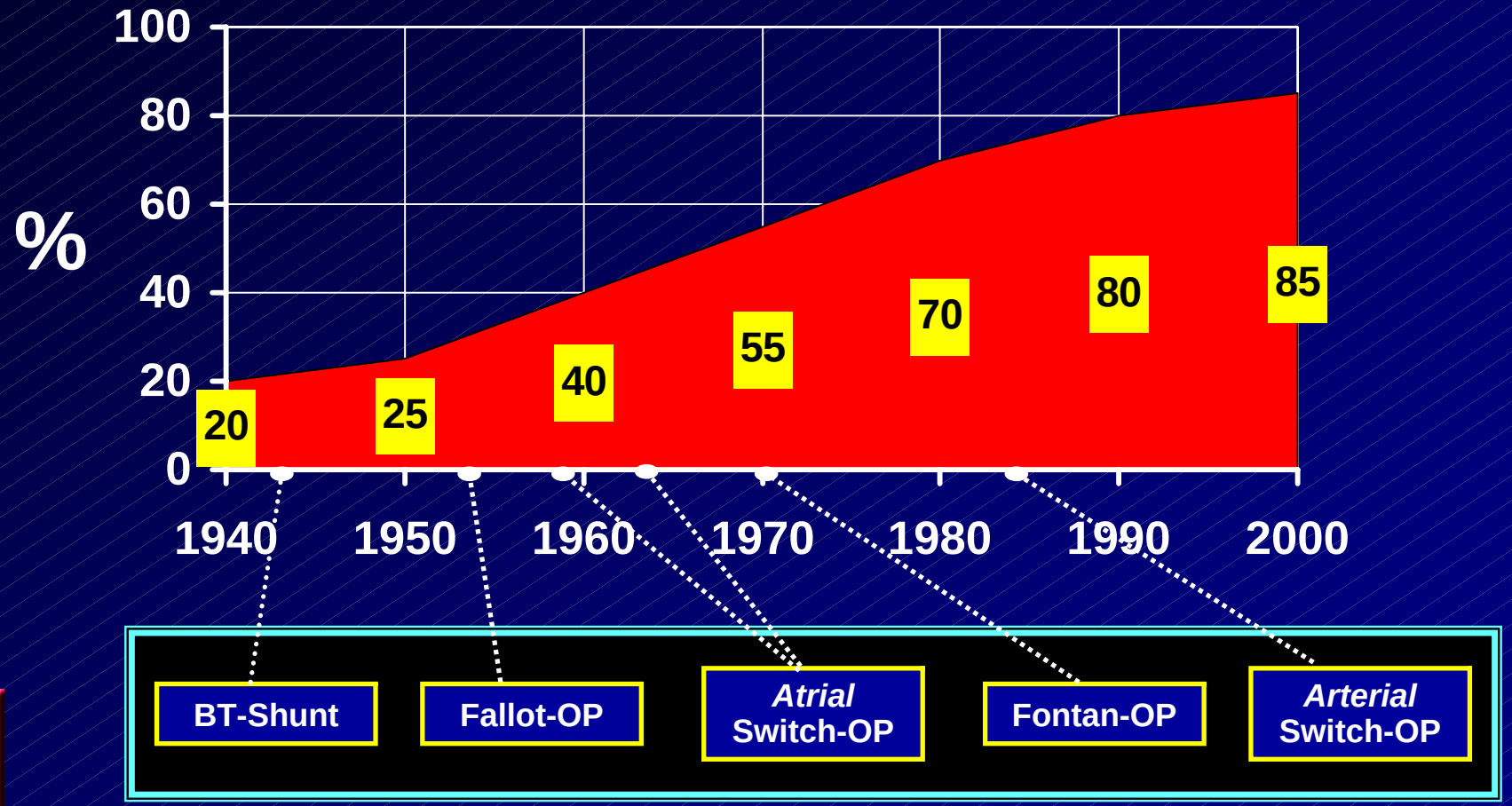
Survival of Complete TGA



Toronto

Courtesy by Dr. W. Williams

Survival Rate in CHD and Milestones in Cardiac Surgery



Toronto

Courtesy of Dr. H. Kaemmerer, Munich

Outline

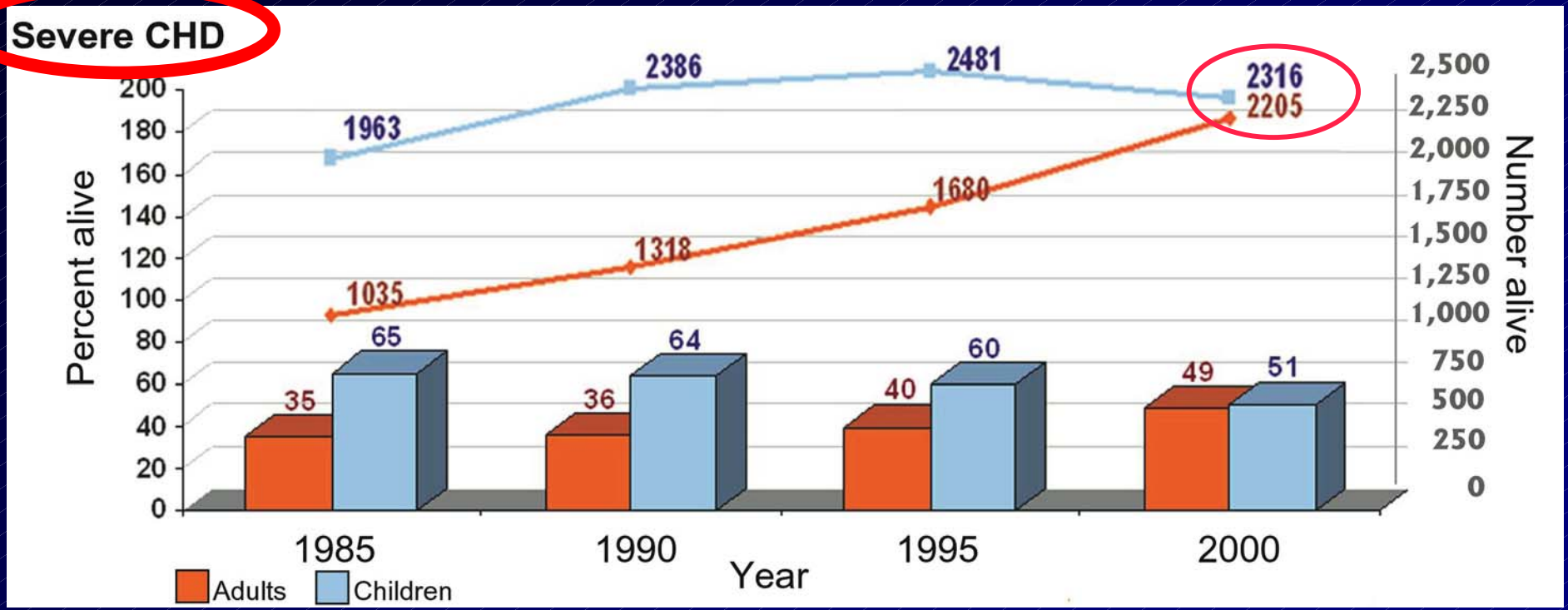
- History
- The adult population
- Do we cure CHD patients?
- Summary

Prevalence of Severe and Other CHD

	Adults Alive in 2000		Children Alive in 2000	
	n (%)	Prevalence per 1000 Adults	n (%)	Prevalence per 1000 Children
All congenital heart lesions*	23 563 (100)	4.09	18 979 (100)	11.89
Severe lesions				
TOF or truncus arteriosus	1001	0.17	778	0.49
AVCD	834	0.14	914	0.57
Transposition complex	235	0.04	424	0.27
Univentricular hearts	150	0.03	213	0.13
All severe lesions	2205 (9)	0.38	2316 (12)	1.45

Marelli AJ, et al. Circulation 2007; 115: 163-172

Changing Age Distribution of Severe CHD 1985 - 2000



Toronto

Marelli AJ, et al. Circulation 2007; 115: 163-172

Extrapolation: Quebec Data

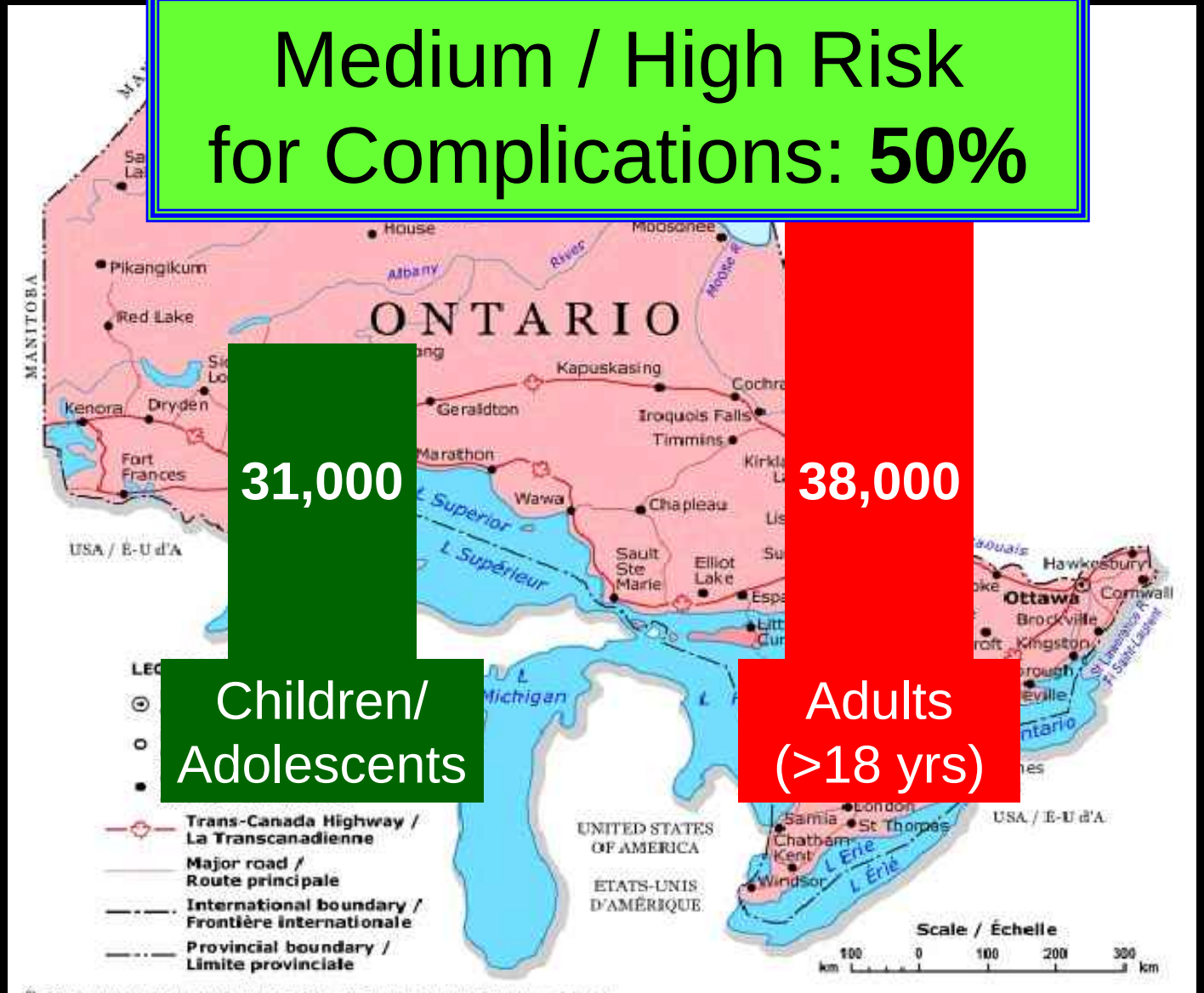
Annual Increase ~ 1,000 Adults per Year

Prevalence of Adults with CHD
(4.09 per 1000)
~ **96,000**

Prevalence of **severe CHD** in Adults
(0.38 per 1000)
~ **9,000**

CHD Patients in Ontario

Medium / High Risk
for Complications: **50%**



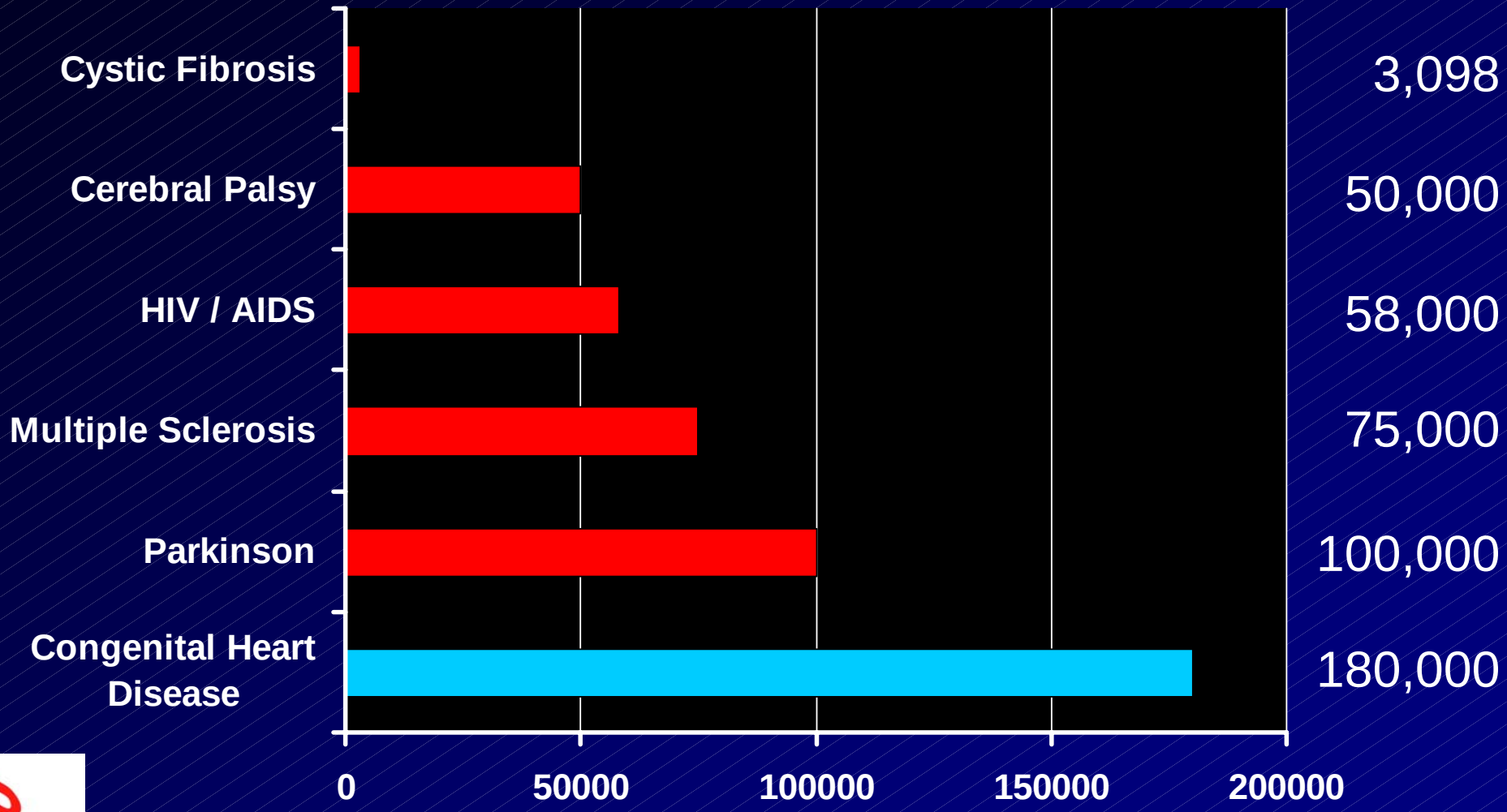
31,000

Children/
Adolescents

38,000

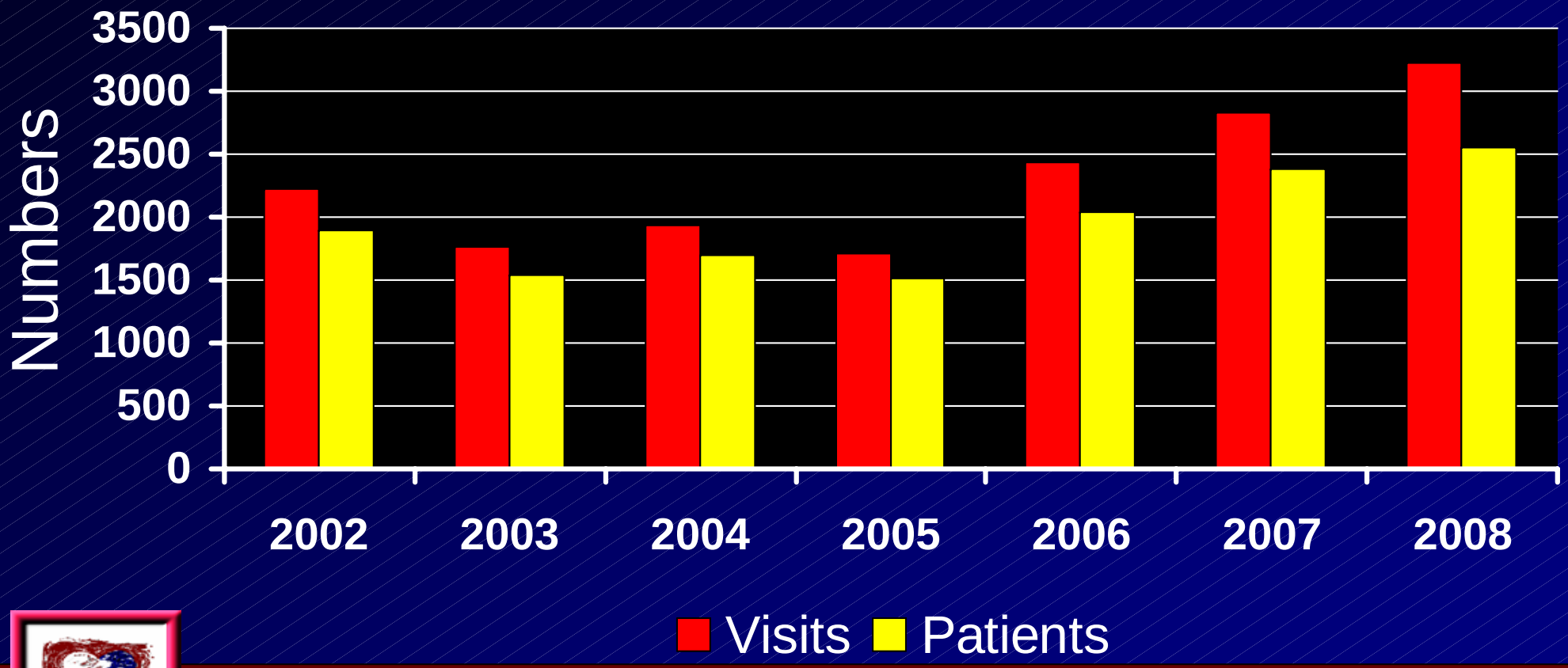
Adults
(>18 yrs)

Canadian Disease Statistics



Sources: Cerebral Palsy Canada, Cystic Fibrosis Association, Public Health Agency of Canada; Canadian Congenital Heart Alliance

Workload in Outpatient Clinic by Year



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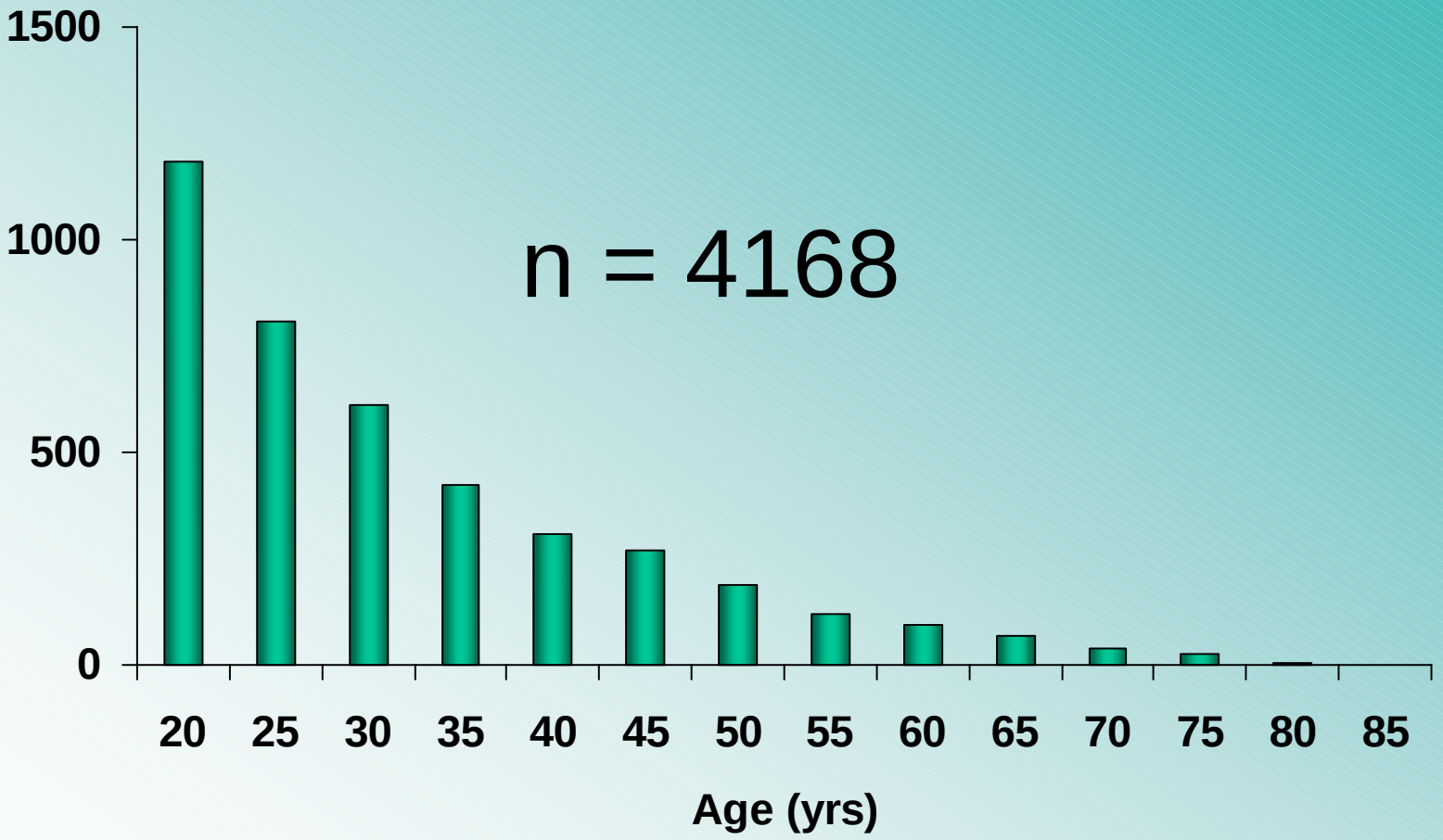
SARS closed the clinic in 2003 for several months

Age Distribution



EUROPEAN SOCIETY OF CARDIOLOGY

Heterogenous Population

- Diagnostic groups
  - Shunt lesions
  - Transposition complexes, Ebstein
  - RVOTO / LVOTO, etc.
  - Pulmonary hypertension
- Surgical procedures
  - None
  - Palliative shunts
  - Repair

Outline

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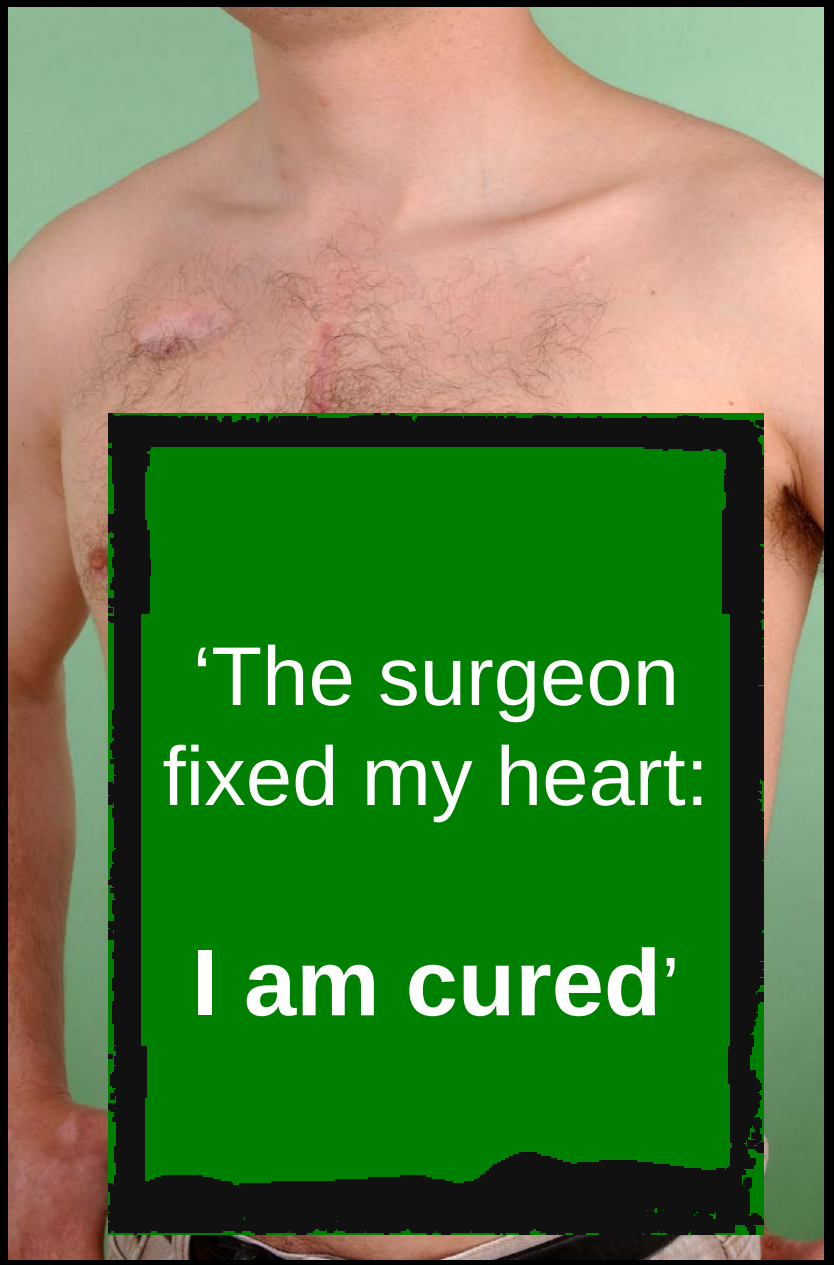
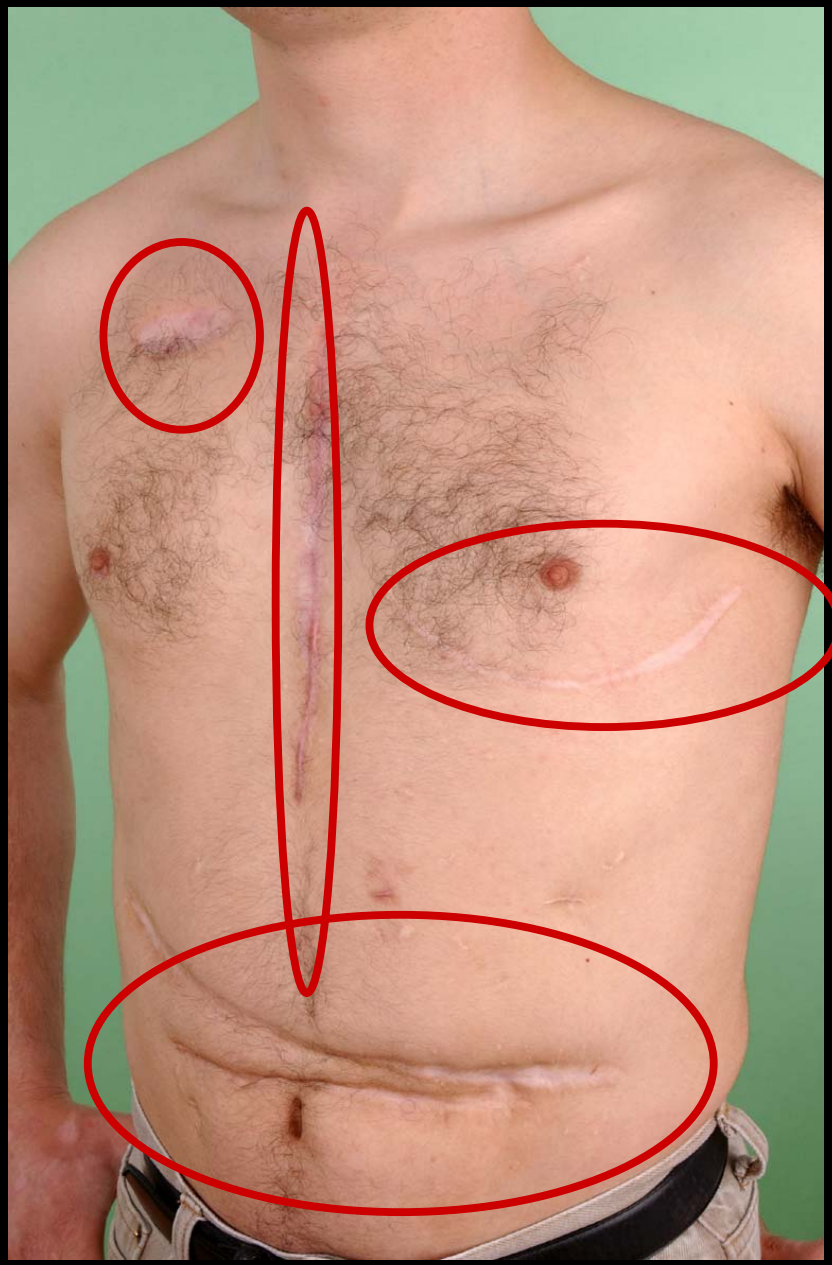
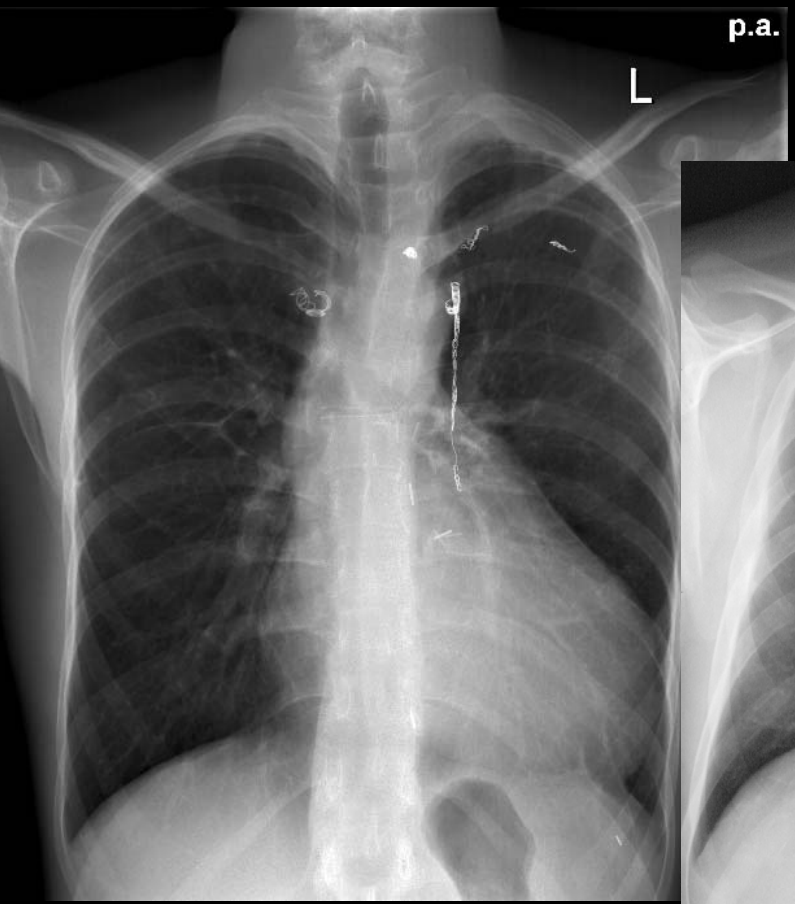
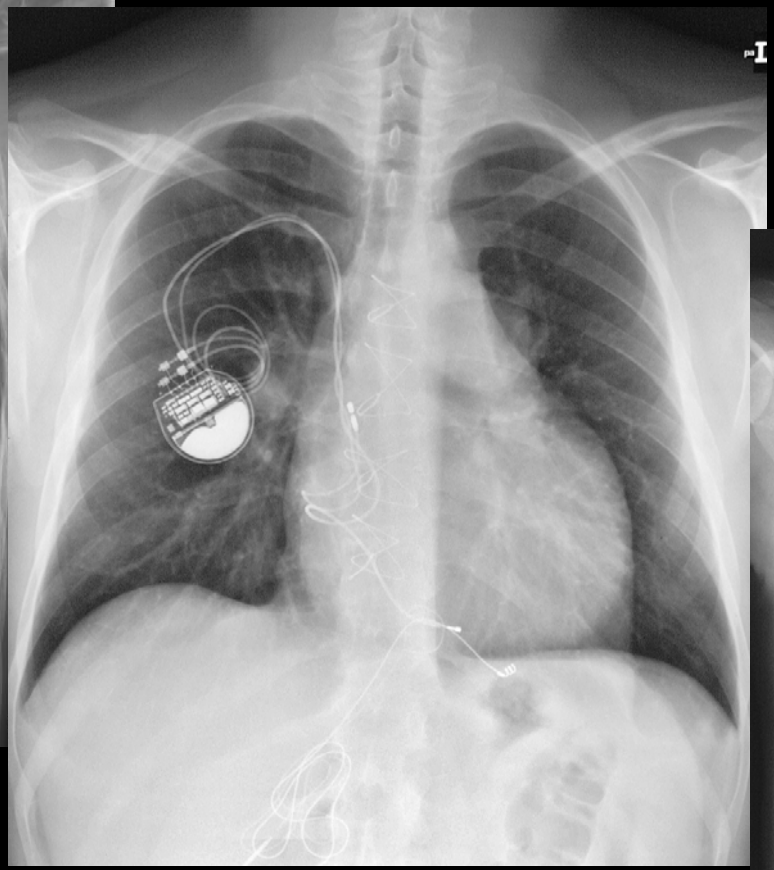
Toronto

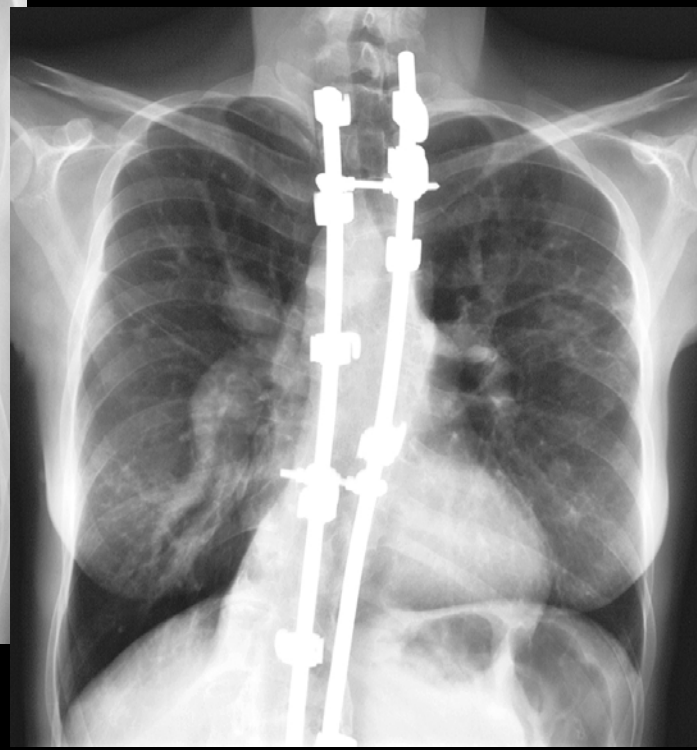

‘The surgeon fixed my heart:
I am cured’



Hardware……







Volume 97, Number 1 January 1989

The Journal of THORACIC AND CARDIOVASCULAR SURGERY

J THORAC CARDIOVASC SURG 1989;97:1-9

Honored Guest's Address

Do we really correct congenital heart defects?

J. Stark, FRCS, FACS, FACC, *London, England*

Surgery is corrective, if.....

- ...ventricular function is normal
- ...life expetancy is normal!
- ...there is no need for therapeutic measures during follow-up!

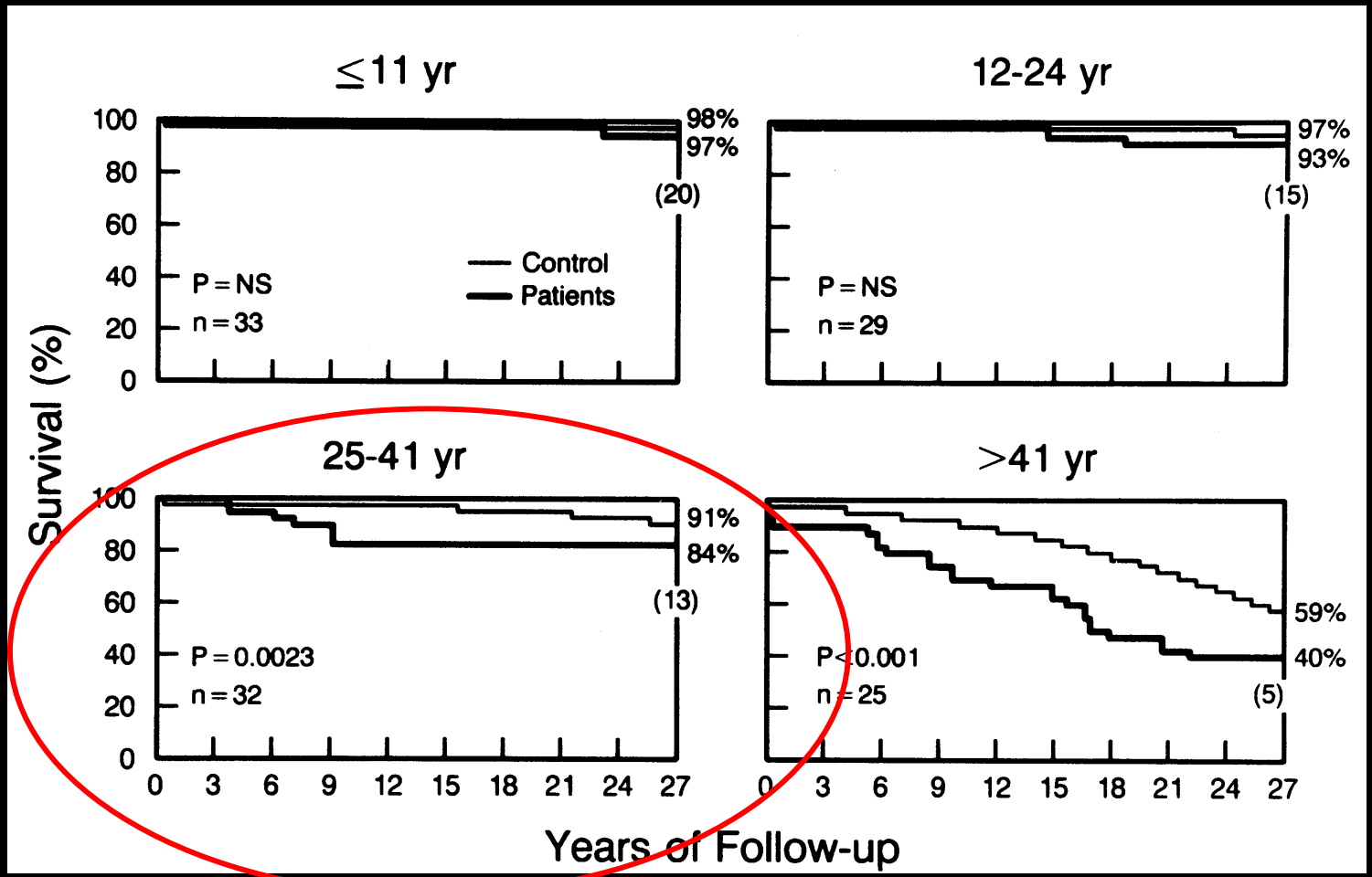
Toronto

Corrective Surgery....

- Atrial septal defect
- Ventricular septal defect
- Patent ductus arteriosus

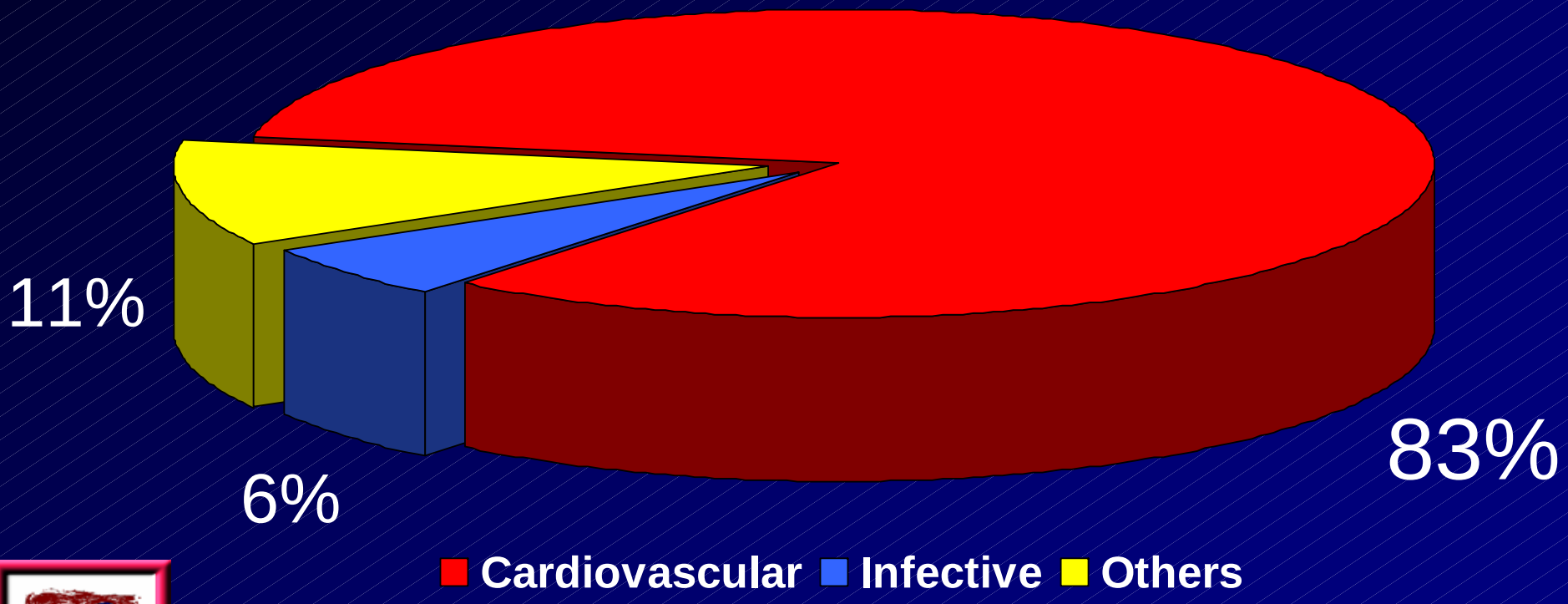
... if treated during childhood!!

Long-Term Survival after ASD Closure by Age



Murphy JG, et al. N Engl J Med 1990;323:1645-50

Emergencies



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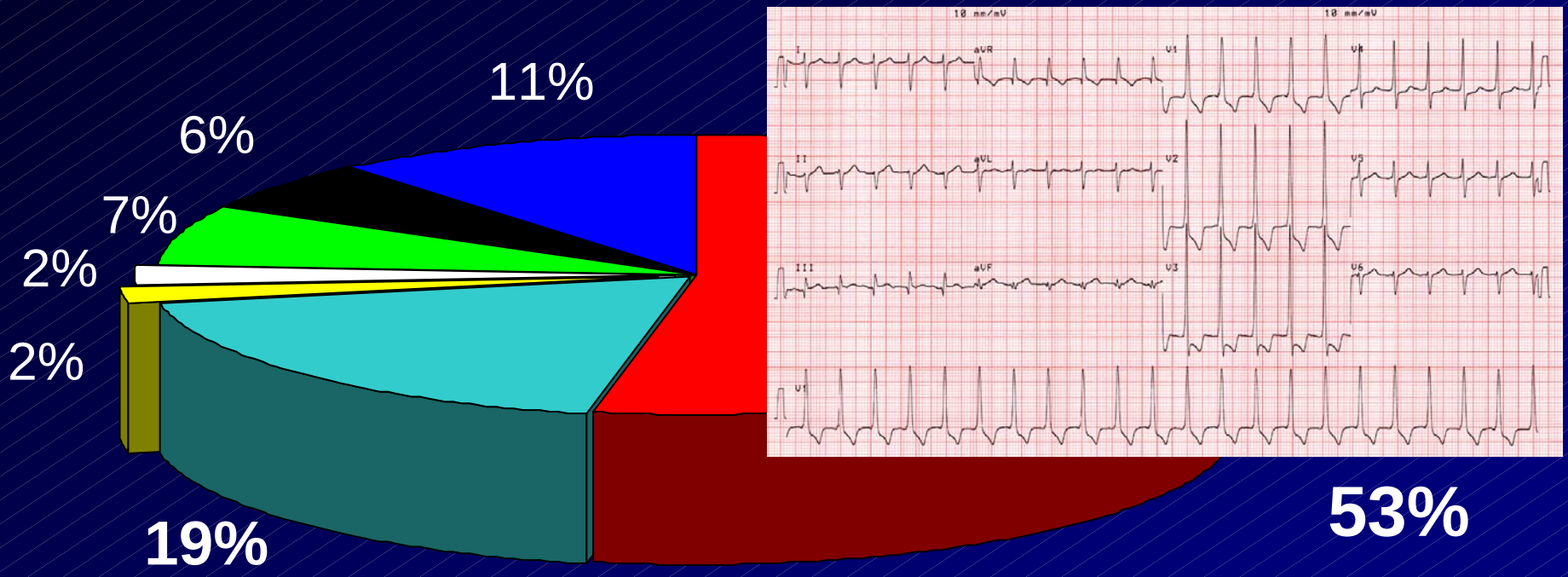
Kaemmerer H, Oechslin E, ... Hess J. J Thorac Cardiovasc Surg 2003; 126:1048-52

Cardiovascular Emergencies



- Arrhythmias
- Heart failure
- Syncope
- Cerebral ischemia
- Other cardiovascular
- Infections
- Others

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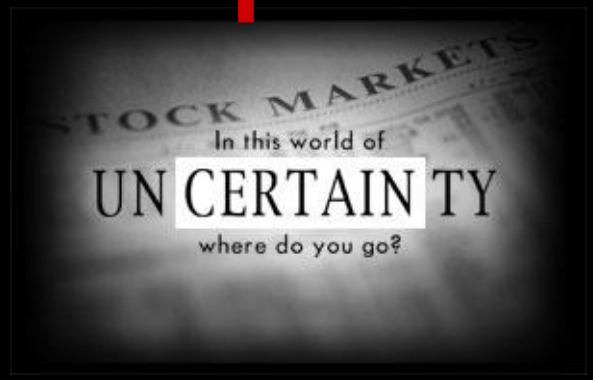
Kaemmerer H, Oechslin E, ... Hess J. J Thorac Cardiovasc Surg 2003; 126:1048-52

Psychological Injury



Long – Term Challenges

- Arrhythmias
  - Supraventricular / ventricular
- Reoperations
  - Failure of conduits / artificial valves
  - Residual shunt / regurgitation / obstruction
- Heart failure
  - Medical therapy / heart / (lung) transplantation
- Biomedical and psychosocial concerns / barriers

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Summary

- The *demographics* of CHD population have changed:
  - Increasing number of adults with CHD
- *Distribution* of CHD:
  - Prevalence of pts with *severe CHD* is increasing more rapidly in adults than in children
  - Equalizing numbers of adults and children with severe CHD

Summary

- *Mortality and morbidity are shifting* away from the young and towards the adult
- *Workload* is increasing

Impact on Health Care System!

Congenital Heart Disease

- Heterogeneous population – the patient profile is changing
- Most pts are not ‚fixed‘
  - Life-long risk for complications
- Expert care with knowledge and expertise in:
  - Anatomy and phsyiology
  - Long-term outcome
- Multi-disciplinary team approach in designated centres

Designated Centre for Pt Care

Patients with ***named conditions or operations*** are **complex**

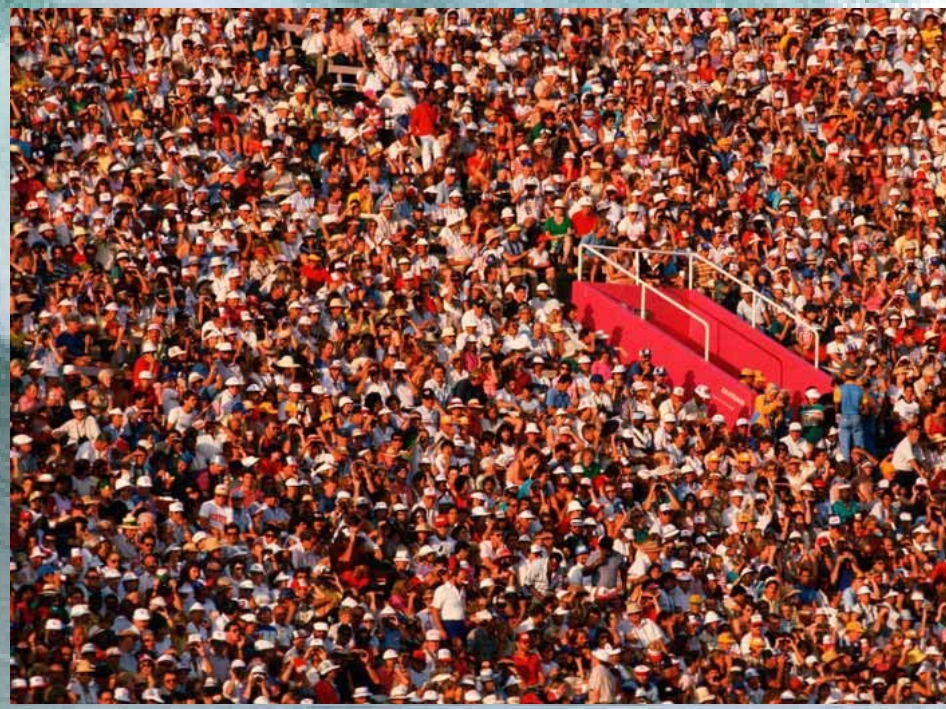
Specialized centre

Toronto

CHD is a Continuum from Fetal Life until Adulthood

ACHD TSUNAMI:
Catch The WAVE!

- Arrhythmias
- Heart Failure
- Pulmonary Hypertension
- Re-Intervention
- Psychosocial Issues
- Reproduction
- Premature Death



ACHD Team 2008